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# THE BACKWARD BABY

A TREATISE ON  
*IDIOCY AND THE ALLIED MENTAL DEFICIENCIES  
IN INFANCY AND EARLY CHILDHOOD*

BY  
HERMAN B. SHEFFIELD, M.D.  
NEW YORK

FELLOW OF THE NEW YORK ACADEMY OF MEDICINE AND THE AM. MED. ASS'N;  
AUTHOR OF MODERN DIAGNOSIS AND TREATMENT OF DISEASES OF CHILDREN,  
PEDIATRIC MEMORANDA, AND THE BABY'S RECORD AND HEALTH,  
AND CO-AUTHOR OF PRACTICAL PEDIATRICS. FORMERLY MEDICAL DIRECTOR,  
BETH DAVID HOSPITAL, INSTRUCTOR IN DISEASES OF CHILDREN,  
NEW YORK POST GRADUATE MEDICAL SCHOOL AND HOSPITAL,  
AND ASSOCIATE BABIES' HOSPITAL (O. P. D.).  
VISITING PHYSICIAN TO THE PHILANTHROPIN HOSPITAL  
AND THE NORTHWESTERN DISPENSARY, ETC.

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*WITH TWENTY-TWO ORIGINAL ILLUSTRATIONS IN THE TEXT*



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## PREFACE

THE aim of this Prize-Essay is to present to the profession a practical survey of the etiology, pathology, diagnosis and treatment of the diverse mental deficiencies as they occur in children under five years of age. The existing excellent monographs and text-books on the subject are almost exclusively devoted to the study of feeble-mindedness in children of school age and adults. It has always appeared to the author, that with a more thorough knowledge of idiocy and the cognate affections in infants, the physician would be very much better prepared to ameliorate, or possibly cure, these conditions before the underlying lesions have permanently destroyed the cerebral functions.

Owing to paucity of literature at his disposal the author was obliged to rely chiefly upon his personal observations. He hopes therefore that his work will be judged graciously should

some of his observations fail fully to coincide with those of keener and more experienced clinicians.

H. B. S.

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# TABLE OF CONTENTS

## CHAPTER I

Nature and Pathogenesis—Heredity—Darwin's Theory, Modified—Modus Operandi of Hereditary Transmission of Amentia—Syphilis—Alcoholism—Consanguinity—Uterine Disease—Involvement of Ductless Glands—Antenatal, Natal and Postnatal Traumatism—Febrile Diseases .....*Page* 1

## CHAPTER II

Examination of the Patient—Family and Personal History—Physical Examination—Stigmata of Degeneration—Normal Mentality and Its Development—Impaired Special Senses—Status Idioticus—Mental Tests.  
*Page* 19

## CHAPTER III

Special Groups of Amentia—Microcephalus—Hydrocephalus—Paralytic Amentia—Amanrotic Family Idiocy—Mongolism—Cretinism—Infantilism—Moramentia—Their Differential Diagnosis.....*Page* 47

## CHAPTER IV

Prophylaxis—Eugenics—Judicious Management of Pregnancy and Labor—Care of the Newly Born and of the Older Child—Mental Training of the Normal Infant .....*Page* 122

## CHAPTER V

Active Treatment of Amentia—General Hygiene—Training by “Incentive” Method and Physical Therapeutic Measures—Organotherapy—Thyroid Transplantation—Drugs—Surgical Procedures . . . . .	<i>Page</i> 137
LITERATURE . . . . .	<i>Page</i> 176
INDEX . . . . .	<i>Page</i> 181

# THE BACKWARD BABY

## CHAPTER I

Nature and Pathogenesis. Heredity. Darwin's Theory, Modified. Modus Operandi of Hereditary Transmission of Amentia. Syphilis. Alcoholism. Consanguinity. Uterine Disease. Involvement of Ductless Glands. Antenatal, Natal and Postnatal Traumatism. Febrile Diseases.

AMENTIA is not an affection *sui generis*, a precise morbid entity, but merely a symptom of a large group of congenital and acquired pathologic conditions, principally of the brain and the ductless glands. The degree of mental debility is very variable and not rarely incommensurate with the extent and gravity of the causal organic lesion. Thus, profound idiocy is frequently encountered with seemingly insignificant structural changes in the brain or elsewhere; and *vice versa*, gross brain lesions may occasionally be accompanied by only slight feeble-mindedness. As a rule, however, definite postmortem findings, with predominance of characteristic lesions in certain types

of cases, are observed in the great majority of cases of amentia, so much so, as to permit—in accord with the underlying pathologic anatomy—the classification of idiocy and the allied mental deficiencies into distinct groups (e.g., idiocy with microcephalus, hydrocephalus, athyrosis, etc.), which will receive due consideration in another chapter.

In almost all forms of amentia the cerebral convolutions are more or less modified and irregular in outline and diminished in number. They are either agglutinated or separated by widely gaping grooves. Frequently there is an appreciable difference in the size, shape and weight of the hemispheres, or an asymmetry of the corpora striata, the peduncles, or the pyramids, or even the absence of one or more of these bodies. Similar changes also are often observed in the cerebellum, and occasionally in the pons, medulla and spinal cord.

Microscopically, we can readily detect an arrest of development or disease of the nerve cells of the brain cortex, of the nerve fibres, and of the neuroglia. The nerve cells are immature, irregularly arranged or numerically deficient.



The nerve fibres are greatly diminished in number, more especially in the frontal and parietal lobes, which seem most concerned with the highest intellectual functions. The neuroglia is quite frequently sclerosed either in certain portions of the brain or in its entirety.

Sclerosis and porencephaly usually predominate among the lesions encountered in the different varieties of amentia of infancy; occasionally, however, neoplasms, especially cysts, and local softening are detected postmortem in cases in which they were least suspected during life.

The cerebrospinal fluid is either increased or diminished in quantity, depending chiefly upon the size of the skull and the amount of brain structure within it.

In addition to the diverse pathologic alterations in the central nerve system, postmortem examination of mental defectives invariably discloses also several lesions in other parts of the body. The ductless glands, more particularly the thyroid, thymus, pituitary and adrenals, are often in a state of rudimentary development or degeneration. The cranial bones are either unusually thick or thin, and the diploë is diminished. The tubular bones are thick and short

and often deformed. It is not uncommon to find congenital anomalies of the heart and blood-vessels, and of the abdominal organs, as also malformations of the eyes, ears, palate, fingers and toes. Indeed, these anomalies are so prevalent, that they are generally accepted as special "stigmata of degeneration" (see p. 25) and of great diagnostic importance.

Contemporary authorities are very much inclined to advance heredity to the forefront of the etiologic factors of mental degeneracy, Tredgold, for example, going as far as to claim a neuropathic ancestry in from 60 to 70 per cent. of the cases of amentia. While this estimate may precisely agree with the histories of aments housed in asylums and special hospitals for idiots, it seems to me that these percentages by far exceed those obtained in private practice. Statistics in this direction in order to be correct would have to embrace not only the personal and family histories of the institutional cases (who almost invariably are of the worst stock and lowest class of society, and whose very life and environment are conducive to mental degeneracy), but also of the even larger number of aments who are quietly kept at home, and whose

mental degeneracy is the result of either antenatal, natal or postnatal traumatism or disease, and who often succumb at an early age, not rarely long before the exact state of their mentality has at all been determined.

Those who claim the preponderance of a tainted heredity as the primary cause of mental deficiency *a priori* concur with the views of Darwin and his disciples, who hold that the offspring inherit the essential characteristics of their ancestors. Now, while this doctrine unquestionably applies to the animal species as a whole and to the transmission of normal racial characteristics, I very much doubt that it conforms to the phenomena of disease, which in contrast to normal attributes of the human species form abnormal, unnatural, nay, often merely accidental accessions.

In order to obtain a clear conception of the workings of heredity, I think it best to assume two distinct phases thereof, viz., one permanent, which has become fixed during the long course of evolution; the other, temporary, accidental or transient.

To the first, *permanent*, category belongs the phenomenon of hereditary transmission of nor-

mal racial characteristics. Taking the African negro, for example, we find that irrespective of the laws of variation and mutation his offspring always maintain their racial characteristics, as long as the negro mates with members of his own clan. And even were he persistently to intermarry with descendants of white races, there still would be little likelihood of his offspring ever entirely overcoming the attributes of their African ancestry. It certainly would require a great many generations to swamp the negro individuality, were it at all conceivable that the law of reversion would cease its vigilance and tolerate such an unnatural process of evolution. This, then, represents the permanent phase of heredity. The second, *temporary*, principle of heredity here suggested is strikingly illustrated by the transiency of certain bodily physical anomalies, as for example, supernumerary fingers and toes. These malformations are occasionally observed in several members of one family. But we usually note, that just as soon as these affected individuals intermarry with normally developed individuals, the aforementioned structural anomalies with but very few exceptions promptly disappear in their future generations, for the very good rea-

son that supernumerary fingers and toes are useless, abnormal and unnatural acquisitions, and hence are dropped by nature at the earliest opportunity. This phase of temporary heredity applies with equal force to anomalies of development of the central nerve system. Indeed, so anxious is nature to eliminate anomalies of development, be they physical or mental, that the great majority of degenerates are destroyed in the germinal, embryonic, foetal, or early post-natal stages of life, or if they happen to survive, are usually rendered sterile, in order to prevent the procreation of their kind.

With these considerations in view I cannot help but hesitate to believe that heredity really plays so important a rôle in the propagation of mental deficiencies as is generally supposed and am inclined to place much more responsibility upon acquired etiologic factors. This reasoning is partly corroborated by the investigations of Scholomowitch, Keller and Diem who found that the difference in the degeneracy-ratio among the offspring of sane and insane ancestry is only about ten per cent. in favor of the former.

The fact frequently cited that on rare occasions (e.g., the famous, or rather notorious

Jukes' family) we do meet hereditary mental degeneracy in several generations, does not in the least controvert the here proposed modification of Darwin's theory. In fact, in a way it even confirms it, since it can readily be shown that, as a rule, mental degenerates persistently mate with individuals of similar mental caliber (for anyone with sound mind could hardly be induced to mate with an idiot!), and therefore the continuity of intermarriage among defectives generates the phase of permanent heredity previously spoken of—in other words, a new race, as it were, with mentally deficient characteristics, is created which does for some time and would forever transmit its degeneracy to its offspring, were it not exterminated by nature in accordance with the law of natural selection and destruction of the unfit.

The *modus operandi* of hereditary transmission is still veiled in deep mystery. It is generally assumed that in mental degenerates the germ-plasm of the male or female, or of both, is defective, either in the number of its component cells, or in their strength and shape, and in consequence fails to form the impetus essential to

normal development of the brain. It is further postulated that under certain as yet mysterious conditions the germ-cells of the opposite sexes, at the time of their fusion, are capable to influence each other, either for good or bad, in accordance with the laws of natural selection. In all probability a tainted germ-plasm is deficient in more than one of its elements, since the degenerate brings into the world not only a deteriorated brain but quite frequently also several anomalies of other parts of the body, e.g., abnormal heart, extremities, etc. Furthermore, there is ample reason for the belief that the anteconceptional deficiencies in the germ-plasm which are productive of amentia in the child may be the result not only of neuropathy in the parents but also of other pathologic states, more especially tuberculosis, cancer, syphilis, and the like, the toxins of which acting as poisoning and deteriorating agents upon the germ cells, the embryo and foetus and arresting their normal development. Statistics are greatly at variance as to the exact number of the feebleminded children procreated by parents thus afflicted. The family histories obtained are almost always inaccurate, since but very few parents are willing to admit or are

aware of the prevalence of latent tuberculosis syphilis, etc., amongst them. Moreover, it is only with the evolution of Wassermann's, and the tuberculin reactions that the statistics pertaining to the causes of feeble-mindedness have become at all reliable.

In a series of 2,380 cases of amentia, G. S. Shuttleworth found a family history of tuberculosis in 28.31 per cent.; of mental degeneracy in 21.38 per cent.; of epilepsy and allied neuroses in 20 per cent.; of parental intemperance in 16.38 per cent., and of inherited syphilis in 1.17 per cent. This extremely low percentage of syphilis as an etiologic factor of amentia was obtained prior to the employment of the Wassermann-Noguchi test and clearly demonstrates the unreliability of the usual family histories as related by patients or their sponsors. For with the application of the test, the figures assume an entirely different aspect. Thus Lipmann applied this test in a large group of idiots and found it positive in from 9 to 13 per cent. of cases, and among 204 cases of low grade idiocy at the New York Children's Hospital and Schools, C. H. Atwood obtained a positive reaction in 14.7 per



cent. Indeed, with the recently improved methods of detection of the spirochæta pallida in diseased brains, syphilis will be found responsible for by a far greater number of cases of amnesia than heretofore even surmised.

All observers agree that parental alcoholism forms a most potent predisposing cause of mental degeneracy in the offspring. In 1901 the New York Academy of Medicine undertook a careful investigation of the effect of parental intemperance upon their children. The family history of 3,711 school children through three generations was traced with considerable detail, and it was found that the children of temperate parents exceeded in proficiency those of heavy drinkers by about 70 per cent., and that a large number of the encumbered children were mentally deficient to a very high degree. Bezzola reports that out of 70 idiots under his observation 35 were conceived during the wine harvest of fourteen weeks during which the Swiss earouse, while the other 35 idiots were conceived during the remainder of the year. Leppich claims that out of 97 children conceived at the time when one or both parents were intoxicated

only 14 were born without noticeable mental or physical defect. L. Mervin Maus presents the following very enlightening table of the relative proportion of degeneracy among the inhabitants of Prohibition States and of those in which the sale of liquors is not restricted, which, if correct, certainly speaks volumes for National Prohibition as an efficient means of prevention of mental degeneracy:

Kansas .....	1 to 873
Indiana .....	1 to 609
Maine .....	1 to 590
New Hampshire .....	1 to 473
Ohio .....	1 to 449
Illinois .....	1 to 473
Rhode Island .....	1 to 436
Michigan .....	1 to 419
Wisconsin .....	1 to 376
Virginia .....	1 to 375
Connecticut .....	1 to 311
New York .....	1 to 290

The etiologic relation of consanguinity to amentia is still subject to controversy. It undoubtedly greatly depends upon the physical and mental condition of the individuals concerned. However, it has often been observed

that all hereditary predispositions to disease in the parents are markedly intensified in the offspring. Deafmutism is particularly prone to occur as a result of union of near relatives.

The postconceptional causes of mental deficiency acting upon the embryo and fœtus are as prolific as and possibly more so than those exerting their influence through heredity. Notwithstanding the purity and normal activity of the parental germ-plasm, it may yet fail in its destiny, if the soil in which the seed is to grow is lacking in the essential prerequisites for healthy growth and development. Let me enumerate the various intrauterine morbid conditions which tend to disturb the normal progress of the embryo or fœtus—in one case, e.g., acting harmfully upon the extremities or heart, in another upon the central nerve system.

1. Disease of the uterine tissues surrounding the impregnated ovum preventing uniform contact between the maternal and embryonic structures and facile absorption of nutriment, viz.: Endometritis, atrophy or hypertrophy of the serotina and reflexa, anomalies of the placenta, either in size or posture, or degenerative changes

therein, such as hemorrhages, white infarctions, fatty, calcareous and pigment deposits and cysts, all of which tend to produce extensive destruction of placental tissue and thus to curtail the blood and food supply of the fœtus.

2. Internal or external violence acting either directly or indirectly upon the fœtus.

The internal use of oxytocics and drastic cathartics to abort, or attempted interference with pregnancy by means of local irritants, instruments or other mechanical contrivances; severe falls, blows, etc., upon the abdomen have from time immemorial been accepted as potent factors in the arrest of normal fœtal development.

3. Excessive intra- or extra-uterine pressure hampering the commodious and equable expansion of the rapidly growing fœtus, thus:

Polyhydramnios, and other abnormalities of the amnion;\* twin birth of large size; large intra- or extra-uterine neoplasms; maternal dwarf-

---

\* In advancing amnion pressure resulting from smallness of the amnion as a most probable cause of achondroplasia, Dr. Murk Jansen suggests that smallness of the amnion may produce other phenomena of pathologic embryology besides that of achondroplasia. Sometimes the vertebral column is curved and the development of the brain is disturbed, and amnion pressure, if exerted during the first and second weeks, is able to cause wrinkling of the embryonic axis, with destruction of ecto- and mesodermal elements (anencephaly).

ism, with marked pelvic deformity, may mechanically interfere with normal development of the foetal vital organs.

4. High degrees of toxemia from febrile affections or poisoning from slow morbid metabolic processes, e.g., typhoid, tuberculosis, and diabetes, especially during the early period of pregnancy may greatly affect the foetus; and, finally, serious domestic trouble, grave mental anxiety and extreme fright with prolonged agitation during the early stages of pregnancy may so undermine the general health of the mother as to indirectly disturb the normal processes of growth and mental development.

In this connection it is not amiss to emphasize that many of the dystrophies, especially of the brain, not rarely observed in prematurely born infants, are the direct or indirect result of some microscopic or gross pathologic changes either in the thyroid, parathyroids, thymus, adrenals or the hypophysis originating at an early period of intrauterine life.

There still remains another large group of mentally deficient infants who though apparent-

ly normal until birth, show definite manifestations of amentia some time thereafter. Traumatism during delivery has always been recognized as a highly potent factor in the production of idiocy and the allied mental deficiencies, the statistics relative to these cases ranging anywhere between 15 and 30 per cent. Where the cranial bones are fully developed and the maternal pelvis is free from extreme contraction or deformity, it is doubtful whether tedious labor *per se* is responsible for mental deficiency developing during early childhood. On the other hand, forcible instrumental delivery of a soft skull impacted in a narrow rickety pelvis is bound to effect some injury to the brain and leave behind a permanent mental deficiency in the child, more especially if the parietal and frontal lobes sustain the brunt of the injury. Occasionally severe asphyxia neonatorum is traceable as an immediate cause of amentia, undoubtedly owing to suggillation and quite severe hemorrhage of the meninges and even of the brain that often accompany prolonged asphyxia. Amentia following natal traumatism not rarely makes its appearance several months or years after the injury has been received, and is often preceded by epi-

leptic convulsions which are attributed to all sorts of immaterial causes. Traumatism in early infancy is an especially frequent cause of mental degeneracy in children of the slums, whose parents, either for want of means or good sense, are very apt to leave their small children to shift for themselves, so that knocks, falls and bruises form part and parcel of the miserable lot of their unfortunate babies. Apparently "the Lord takes care of the helpless children," for were it otherwise the hordes of idiots would have swelled beyond calculation or imagination.

Febrile affections, more particularly meningitis, encephalitis and exanthemata, form very material etiologic factors of permanent degeneration of the infantile central nerve system. Acquired hydrocephalus supervening grave gastroenteric intoxication, severe attacks of pertussis (by inducing cerebral hyperemia or local hemorrhage), and acquired diseases of the thyroid (e.g., endemic cretinism) most probably stand next in frequency as etiologic factors of amentia. Not rarely also mental backwardness is traceable to deprivation of senses, such as vision and hearing, particularly if these unfortunate children are

not given the benefit of expert treatment and training. Several authors mention malnutrition, rachitis and adenoids as rampant causes of mental deficiency in young children. The mental dulness in these cases, however, is only temporary, promptly giving way to full vigor upon removal of underlying mentally retarding factors.



## CHAPTER II

Examination of the Patient. Family and Personal History.  
Physical Examination. Stigmata of Degeneration.  
Normal Mentality and Its Development. Impaired  
Special Senses. Status Idioticus. Mental Tests.

AFTER reviewing the aforementioned intricate causes of amentia we can readily appreciate the importance of obtaining a clear personal and family history of the case in question. It is especially essential to learn whether the amentia is congenital or acquired, since it furnishes the most reliable clue to the prognosis and treatment of the case. In taking the history, however, it is almost equally important to remember that histories obtained from parents are not always very reliable; firstly, because the latter are rarely very certain of their own mental shortcomings, and more especially of those of their ancestors; secondly, they are usually loath to admit degeneracy in their immediate family; and, thirdly, either for want of good judgment, or in the hope of favorably to influence the doctor's opinion, they are very apt to conceal certain mental inferiorities of their infants or to exaggerate their mental

powers and thus to mislead the examiner. However unreliable the history may be, it always furnishes at least a few threads of information which help to direct our attention to some mute points in the diagnosis, which otherwise would escape our observation. Let me cite an interesting case in point. About two years ago I had occasion to pronounce an eighteen-months-old infant as mentally deficient (Mongolism), as a result of antenatal arrest of development, though the history of tedious labor with low forceps delivery; compression of the right parietal bone; softness of the entire skull, and marked separation of the sagittal and coronal sutures, and general physical debility, led several physicians to attribute the mental backwardness to rachitis and possibly also to localized cerebral traumatism during birth. In taking the history of the case I elicited the facts that the baby's mother had a very severe "nervous breakdown" before marriage and that her mother had for over a year been suffering from melancholia during menopause. Further, that the mother had previous to this child given birth to a frightful monstrosity. On examination I found the patient's ears and fingers markedly deformed, the tongue

large and protruding, the teeth irregularly developed and implanted, and her mentality practically that of a three-months-old infant. There could be no question about the diagnosis of idiocy, still the mother was reluctant to accept this diagnosis and urged me to refer her to a clinician of vast experience for confirmation of my opinion. The following week I received a letter from this learned gentleman outlining the mode of treatment of the case which "judging by the large open fontanelles and extreme debility is undoubtedly suffering from marked rachitis." I courtly replied that I was not very particularly interested in her rickets, but expected to obtain his authoritative opinion as regards her mentality. He promptly apologized for his oversight, explaining that the mother had consulted him in reference to the child's general debility, that the baby was crying during his examination, hence offered no opportunity to observe her facial features, and that he failed to inquire into the history of the case, because the symptoms so strongly pointed to rachitis.

The taking of the family history should include questions as to insanity, idiocy, dipsomania, syphilis, tuberculosis, cancer, epilepsy and mon-

strosity in the immediate family, both on the father's and mother's side. The condition of the previously born children, if any, at the time of birth and thereafter. Diseases of the mother immediately before and during pregnancy. The mental state of the mother during pregnancy, especially as regards grief, fright or extreme emotion from other causes. Traumatism during pregnancy, possible means used to abort, drug habits, etc.

The past and immediate history of the patient should furnish us with all details as regards asphyxia, bleeding from nose and mouth and injuries during labor; the appearance of the head and other portions of the body immediately after birth; convulsions at this time or at any time thereafter; mode of feeding and physical progress of the child; diseases it suffered from, particularly as to exanthemata, pertussis and otitis; traumatism and its immediate consequences; the period at which the infant was able to hold its head erect, to sit up, to stand and walk, and the teeth made their appearance; and also the age of the baby when it made the first attempt to speak. It is also advisable to let the mother relate in her own way what she observed of the

mental acumen of her child, more especially in reference to its progressive or regressive character. This is important, as we intend to show later that some idiots, e.g., cretins and amaurotics, get more stupid as they grow older.

Lest we forget, let me state right now that while the parents are busy relating their "experience" and responding to questions, and the patient is still in a passive mood, unmolested and unaroused by the ordeal of the physical and mental examination, the physician should avail himself of the opportunity to note the attitude and behavior of both the parents and the child and to size up the general aspect of the case. Indeed, as with increased experience we gradually learn to see aright, it is often surprising how easily we can arrive at a correct diagnosis by mere superficial observation of the patient. This statement is not intended to convey the idea that such momentary examinations should suffice to express a positive diagnosis. Quite the contrary, irrespective of what impressions we gain at a glance, we must never omit a very careful and minute physical examination of the child and, this completed, to apply all the mental tests presently

to be outlined. But I do desire to lay especial stress upon the importance of training ourselves to see a great many things at a glance. After undressing the child we proceed with the usual physical examination of children, but devote a little more attention to inspection and mensuration\*

Age	Weight lbs.	Height in.	Circumference of		
			Head in.	Chest in.	Abdomen in.
1 Month	8	21.75	13.75	13.50	13.50
2 Months	10½	23.25	15.40	14.09	14.09
3 Months	12	24.00	15.80	14.70	14.70
4 Months	14	24.75	16.14	15.30	15.30
5 Months	14¾	25.21	16.60	15.88	15.88
6 Months	15½	25.75	17.00	16.07	16.07
7 Months	16¼	26.00	17.16	16.90	16.75
8 Months	17	26.50	17.37	17.00	17.00
9 Months	17¾	26.75	17.50	17.25	17.25
10 Months	18½	27.25	17.66	17.50	17.50
11 Months	19¼	27.75	17.82	17.74	17.75
12 Months	20	29.00	18.00	18.00	18.00
14 Months	21	29.00	18.16	18.16	18.16
16 Months	22¾	29.50	18.33	18.33	18.33
18 Months	23½	30.00	18.50	18.50	18.50
20 Months	24	30.50	18.62	18.62	18.62
22 Months	24½	31.00	18.83	18.83	18.83
24 Months	25	31.50	19.00	19.00	19.00
28 Months	27	33.00	19.16	19.33	19.16
32 Months	29	34.00	19.33	19.66	19.33
36 Months	31	35.00	19.50	20.00	19.50
3½ Years	33	36.50	19.71	20.50	19.71
4 Years	35	38.00	20.00	21.00	20.00
4½ Years	38	38.50	20.21	21.21	20.20
5 Years	41	41.50	20.50	21.50	20.00

which enable us to reveal the pathognomonic signs of amentia, more particularly the Stigmata of Degeneration, which are invaluable in the differential diagnosis between congenital and acquired amentia, and should invariably receive careful consideration. Therefore let us briefly enumerate them, viz.:

#### STIGMATA OF DEGENERATION

1. Abnormalities in size and shape of the head. Softness or bossing of the cranial bones. Marked gaping or premature closure of the fontanelles and sutures. Undue distention or sinking of the fontanelles.

2. Malformations of the ears. Irregularity in size. Undue prominenee or flattening. Misshaped helix, antihelix, tragus, antitragus and lobule. Supernumerary auricles, auricular appendages or atresia auris.

3. Anomalies of the eyes or lids. Drooping of one or both eyelids. Epicanthus and palpebral fissures. Congenital cataract, coloboma iridis or idideremia. Micro- or ano-phththalmus. Strabismus and nystagmus.

4. Malformations of the nose. Saddle-shaped, exceptionally small and broad, or unduly large

and prominent. Partial or complete atresia of the posterior nares.

5. Malformations of the face. Undue prominence of the cheek bones with markedly retracted small chin. Clefts of face and lips. V-shaped or high vaulted palate. Enlarged, protruding and often cracked tongue. Irregularly shaped and implanted teeth, deficiency or excess in their number. Inability to bring jaws closely together, owing to irregularity of dental arches, hence constant dribbling of saliva from half-open mouth.

6. Malformations of long bones. Curvatures of the bones of the upper and lower extremities. Supernumerary fingers and toes or deficiency in their number. Syndactylism or fan-shaped distribution. Disproportion in size of legs and arms. Talipes, spina bifida and caudal formations.

7. Umbilical hernia; diastasis recti abdominis. Anomalies of the genitalia: epi- and hypo-spadias. Malformations of the anus and rectum.

Valuable as a tainted history and the existence of stigmata of degeneration are as diagnostic aids in amentia, they are at best only of relative



value in estimating the mental state of the child in question. It is not at all unusual to meet with perfectly normal children who present a neuro-pathic history and several bodily malformations, and *vice versa*, perfect histories and bodies with inferior brains. Furthermore, infants of certain Mongolian races naturally possess the typical Mongolian facies and yet may be fully as intelligent as, and possibly more so, than a child of the purest white race with an unblemished history and anatomy. Hence, before declaring an infant mentally deficient, it is absolutely indispensable to put it through definite physical and mental tests, which reveal the mentality of children of certain ages and permit not only the distinction between the normal and abnormal mentality, but the degree of mental deficiency as well. The importance of such an examination becomes especially evident when we bear in mind the fact that some infants are merely slow in their mental development as a result of diseases or faulty environment, but promptly unfold their mental powers under proper care and treatment.

In order to be able to estimate the mentality of an infant correctly, we must, of course, have a perfectly clear conception of the normal intelli-

gence at different periods of its existence. We shall, therefore, endeavor to depict the normal mentality of the infant, before attempting to outline the mental tests for one who is less gifted.

According to latest investigation a normal baby can hear and see immediately after birth. It feels pain when it is hurt and cries when it is uncomfortable or hungry, and exercises its extremities and the musculature of other portions of the body, if not immobilized by an overabundance of coverings or fancy frocks and frills and bows and strings.

At one month it begins to locate the direction of sound and momentarily to follow a bright light.

At two months it responds to snapping of the fingers, follows bright objects more or less intently, and rejects ill tasting food or drugs.

At three months it holds its head erect, and can turn it steadily from side to side; it smiles when accosted, shows an inclination to grasp bright objects displayed in front of it, and coos when in good humor.

At four months it begins to recognize its mother or nurse, or those who fondle it; manipu-

lates things put in its hand, e.g., a rattle; plays with its fingers, and brings everything to its mouth.

At five months it knows its mother, nurse or father, and puts out its hands to be taken when they approach. When crying from hunger it stops promptly as the food is brought near, and it opens its mouth—ready for the prey.

At six months it is interested in its surroundings; sits up in a chair, with slight support; shows gratification when taken outdoors.

At seven months it recognizes familiar faces from a distance; grasps after objects placed at a short distance; begins to imitate sounds and syllables;\* laughs aloud, and smiles to everybody, and cries when scolded.

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\* N. J. P. Van Baggen (of the Hague, Holland) distinguishes different periods in the development of the infant's speech. In the first period during the first year, the infant utters involuntary sounds, which must be considered as a simple muscular action of the apparatus of speech produced by an unconscious reaction of the numerous stimuli which the child receives from without. Later on the stimuli becoming more intense, reach, through the spinal marrow, the centres of the cortex, and the child begins to feel the muscular movements and to be conscious of them. About the same time, however, the child begins to hear the sounds it produces. Henceforward it feels those sounds as well as it hears them. Both sensations now leave their traces on the cortex of the brain in those parts which are destined for the motor centre of the muscles of articulation and for the centre of hearing. The sensations of feeling and of hearing the word occur simultaneously

At eight months it attempts to stand if held erect or to creep if placed on the floor. It is often able to repeat "mama" or "papa," to clap hands, to shake bye bye and to perform similar little "baby tricks." It understands several words spoken to it and enjoys a game of "peep bo" and the like.

At nine months it knows its name and also turns in the directions of other persons who are accosted. It easily holds and carries its bottle to the mouth; is able to bite off and masticate solid food. If properly trained, it indicates its desire to urinate and defecate.

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and therefore they become united by simultaneous association. The child now begins to imitate the sound it produces itself and soon thereafter it notices the sounds produced by others and it tries to imitate them.

The child now commences to appreciate the conformity between the sounds it hears and the sounds it utters, the imitation becomes more and more complete, and finally syllables and simple words are pronounced. However, this pronouncing of words has not yet any meaning for him. It is only gradually that the association between the heard and pronounced word and the realization of its significance takes place. This association is brought about by the simultaneous hearing of the word and the seeing of the object which the word indicates. Whenever the child sees its doll, the word doll is repeated till at last the child unites the word doll inseparably with the object itself and henceforward the heard word and its characteristic meaning are fixed in the child's memory. The child enters the third period when it begins to use the words which it knows by memory. When it wants its doll it will pronounce the word even without seeing the object or hearing the word pronounced by standers by.

At twelve months it stands alone, or by holding on lightly to a hand or chair, and in the same manner attempts also to walk. It knows the difference between the articles of food it is accustomed to eat. It throws a kiss or actually kisses, when coaxed to do so.

At fifteen months it makes itself thoroughly understood either by signs and motions or baby language. It can point to the nose, eyes and ears, etc. It is interested in picture-books, colors and different toys; can turn pages and scribble with chalk or pencil. It knows the difference between a cat and a dog, and is often able to name them from life or drawings. It can play a toy piano or mouth-harp.

At eighteen months it usually runs about freely and engages in several games, such as throwing balls, marbles and the like. It can imitate all sorts of performances, such as dance, jump, hide, rock a doll, etc. It knows the difference between right and wrong, and obeys or rebels.

At two years it knows exactly what it wants in the way of food or toys, and, as a rule, is able to call for them. It speaks with characteristic gestures; is able to feed itself, and to distinguish manifestations of the weather (snow or rain).

At two and a half years it can make itself, as it were, useful around the house, i.e., do little errands. It begins to ask questions and to "show off." It recognizes different colors, shows constructive ability by making correct use of building blocks, etc.; carries simple tunes and memorizes more or less lengthy nursery rhymes.

At three years it uses the personal pronoun in conversation. It shows an inclination and some ability to dress itself. It can indicate the seat of pain or annoyance. If instructed, it can count up to ten, at least, and spell simple words, or pick several letters of the alphabet.

The physical and mental activity of the child here depicted pertains, of course, to that of average normal intelligence. Some infants excel others in certain capacities, and *vice versa*. It is not at all uncommon, for example, for some babies to walk and to talk at one year of age, or, conversely, barely to begin either at two years or even later, and yet be perfectly normal in every other respect. But we must set before us a standard of the average and not of the exceptional baby, and with due allowance for delay in development as a result of disease or lack of training, compare the physical and mental activity of the

baby under examination with that of the assumed normal standard.

Judging from the foregoing discussion a normal infant is supposed to acquire the power of seeing, hearing, taste and touch when it reaches the first four months of its life; attention, voluntary motion and perception during the second four months; imitation, speech and understanding in the third four months, and gradually, from month to month, to unfold and to strengthen these qualities, so that at the age of about three years it has developed into a real human being intellectually. Let us now attempt to analyze those qualities as they are manifested in mentally deficient children and to suggest workable mental tests to facilitate their early recognition.

*Vision.*—As a rule, idiots gaze vaguely into empty space or irregularly rotate their eyes in all directions. They rarely follow a bright object placed before them and it is almost impossible to fix their attention upon one point for more than a few moments. In testing their power of vision, however, we must assure ourselves of the absence of congenital or acquired obstruction



to vision, e.g., congenital cataract, large staphylomas and the like. Sollier maintains that blindness is encountered in from seven to eight per cent. of idiots. The importance of an early ophthalmoscopic examination of the eyes cannot too strongly be emphasized, since by this means only are we able to detect optic atrophy, symmetrical changes in the maculae, and choroid tubercles, which are often decisive in the diagnosis of amentia of cerebral origin.

*Hearing.*—The sense of hearing is easily tested by starting some sort of a noise (ringing of a bell, clapping of the hands) while the patient is unawares. The ament who hears will ordinarily be startled by the noise, at least momentarily, even though he usually fails to turn in the direction of the noise. Some aments, e.g., amaurotics, are often violently startled by the slightest clapping of the hands. Deafness in connection with amentia is a rare congenital anomaly and almost never forms the sole cause of true mental deficiency (see p. 109).

*Sense of Taste and Smell.*—One of the very earliest signs of amentia is obtuseness or perversion of sense of taste. Aments either chew everything put in their mouths, regardless of its



disgusting taste, or, conversely, spit out the most pleasant delicacies, because of their inability to detect their agreeable taste. They relish quinine as greedily as sugar, or refuse both. This perversion of taste explains why some aments are gluttons and others again barely eat enough to sustain life. The sense of smell is equally affected, but cannot be tested with any degree of exactitude until the child has attained considerable intelligence. Some clinicians record lack of local or reflex response to irritating odors, such as ammonia. In these cases, however, we are most probably dealing with malformations of the nose (e.g., atresia), so that the strong odor does not at all reach the olfactory nerve.

*Sense of Touch, Pain and Temperature.*—Almost all confirmed idiots are insensitive to pain and temperature, hence are frequently seen burnt, bruised and bitten without showing any signs of discomfort. Indeed, some of them delight in mutilating themselves. It is of daily experience to find a mentally deficient child squatting on the floor, bed or chair, rocking to and fro, diligently cracking its fingers or biting its hands, often until they bleed, and rebelling and howling if interrupted in its apparent state

of enjoyment. So characteristic and impressive is this peculiar attitude of the ament that a few years ago I ventured to describe it as the "Sta-

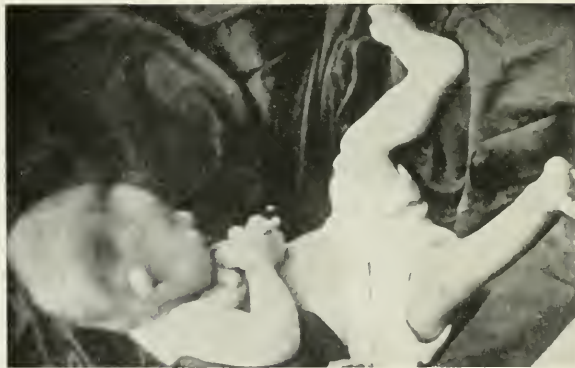
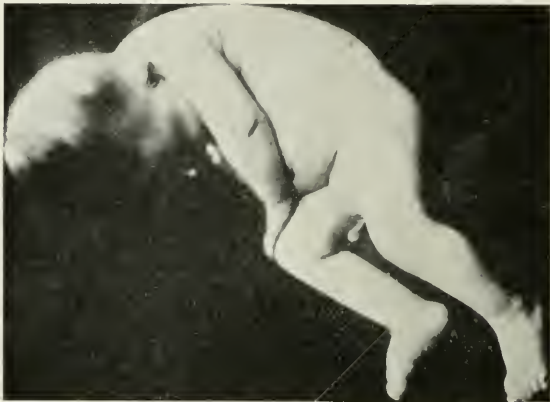


FIG. 1.

tus Idioticus" (see Fig. 1). In congenital amentia there is frequently general anæsthesia, while in acquired cases the anæsthesia is not rarely lo-

calized over large areas of the body, more especially in connection with paralysis. Tactile sense is not nearly as obtuse as that of temperature or pain; in fact, some amients, like the blind, show a distinct hyperacuity of tactile sensibility, by mere touch being able to recognize the individuals who take care of them.

*Attention.*—No other defective mental action so readily betrays the mental incapacity of an infant as his lack of power of attention. A normal infant barely three months old shows its power of attention by turning in the direction of the sound of a bell, for instance, and watches the course of bright objects slowly passed before its eyes. The amient of a much more advanced age, on the other hand, is entirely unconcerned as to what is happening around him. He may suddenly start when frightened by a flash of lightning and he may be aroused from his lethargic state by the approach of one who takes care of him, but he immediately falls back into his hebetude just as soon as the artificial agitation has subsided. It is entirely devoid of initiative and spontaneity, and may for hours sit huddled up in one spot as long as he is not disturbed from sucking his thumbs.

*Perception.*—This utter incapacity of attention, of course, goes hand in hand with dulness of perception. The less attention the ament pays to the doings and actions of others, the fewer are the impressions that reach his brain, and the less capable is his cerebrum to perceive outside impulses. Moreover, his memory is so flighty that he is unable to treasure up for future use the impressions he receives. Again and again, for example, will idiots suffer pain from the effects of burns or other injuries, and yet when exposed to the same or similar harmful forces, they will not at all attempt to guard themselves against injury, for the very reason that from one time to another they forget what happened to them under such circumstances. They rarely recognize familiar faces and cannot differentiate one object from another unless specially trained in this direction.

*Imitation.*—In view of faulty memory, attention and perception, it is hardly to be expected that a degenerate of this sort would be capable of imitation. It is true, some of them do perform little tricks after repeated training, more especially when encouraged by mother or nurse, but their activity is extremely limited, and their

performance very awkward. Unlike normal infants, they do not "show off" spontaneously. Very often after learning one movement they keep on repeating the same almost indefinitely, or until they have managed to learn something else to replace it. The same lack of power of imitation hinders them from engaging in any sort of games, and later in life, to learn to read or write, or to acquire mechanical skill to practice a trade, although exceptionally some aments show considerable constructive talent and ingenuity.

*Voluntary Motion.*—Profound amentia is invariably associated with muscular insufficiency and inco-ordination. Not only are mentally deficient infants lacking in initiative to grasp objects displayed before them, but even if objects are placed in their hands, they are usually incapable of getting a firm hold of or to manipulate them. As a rule, they are unable to measure distance, and hence, like the blind, they feel their way in different directions if they ever manifest a desire to locate a certain object. Amentia is frequently accompanied by paralysis of the extremities, but even in its absence aments very rarely begin to walk before two or three years of

age, chiefly because they are slow to learn the special voluntary movements required in the primary act of walking. In a similar manner they rarely learn to feed themselves with a spoon; they are sure to spill its contents before bringing it to the mouth. Quite a number of aments seem to experience considerable difficulty also to manipulate the tongue, which possibly explains their frequent inability to masticate solid articles of food.

*Speech.*—Marked delay to walk as well as to talk is almost pathognomonic of amentia. Occasionally a mentally deficient infant may succeed to repeat a few single short words at an early age, but it is never able to correctly pronounce several words in succession as to form an intelligible sentence. Some aments as they get older keep on chattering incoherently and without measure, but they are no wiser than others who never utter a single syllable. Hence in judging the mental capacity of the idiot, it is not the number of words he can pronounce that counts, but the way he speaks and what he says. Aments often bring out the words in staccato fashion—slow, broken or “scanning,” and having, as a rule, an imperfect image of words, can-

not pick the right words for the particular things they desire, and therefore fail to make themselves understood.

*Intelligence.*—All the aforementioned attributes of the brain collectively serve to mold the human intellect as a whole. But an infant may be able to see and to hear, smell and taste, pay attention, imitate, perceive outside impressions and, finally, to walk and to emit sounds, and yet not be endowed with normal human intelligence. Practically every domestic animal possesses these faculties. The human mind differs from that of the lower animal by its acquired faculties (not instinct) to distinguish right from wrong, to reason, judge, associate ideas, and to act spontaneously from previous experience. Now, while a properly trained infant, let us say of three years, fully appreciates that it is wrong to get soiled, reasons how best to avoid punishment, e.g., by putting the blame on someone else; associates ideas, by looking for paper when you hand him a pencil; uses judgment, by not attempting to cross the street when seeing an automobile approaching; and finally, shows spontaneity and power of imagination, by making use, for example, of a box, string and cane, to take the place

of a horse, whip and wagon, the mentally deficient child is utterly lacking in all these mental capacities and performs certain actions only automatically by imitations after persistent training. Of course, not all aments are alike in their mental acumen. We must always bear in mind that there are different degrees of amentia, just as there are variously gifted normal infants. But whereas the normal child through outside influences easily and readily acquires certain mental qualities as he gets older in months and years and experience, the abnormal child, owing to some faulty congenital or acquired anomalies of the brain, is unfolding those mental powers at a very much later age, if ever. And it is with the object in view to determine to which period of life the mental capacity of the infant under examination—as compared with the average normal child—corresponds, that we shall presently endeavor to outline helpful mental tests for our guidance.

#### MENTAL TESTS

##### *Mental Age, Six Months*

Move bright object in front of child—note if it follows it.



Ring bell at a distance of about two feet from baby—note if it turns around.

While baby drinks its milk-mixture, remove bottle from its mouth and substitute a bottle containing a trace of quinine, salt or nux vomica, or warm water. Note how it takes any of the solutions. The normal baby shows the possession of sense of taste by promptly refusing even the plain water.

Prick baby's skin lightly with point of needle—watch for prompt facial expression of annoyance.

Hold baby's food at a short distance—watch baby's facial expression of satisfaction and desire to grasp the bottle or breast. Or let mother leave the room and return from another direction; note promptness of attention.

Put baby on mother's lap and note its power to hold its head erect and to sit up with but slight support.

### *Mental Age, Twelve Months\**

While unawares, call infant from a distance—note if it turns in the direction of the voice.

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\* In infants of a year or older it is preferable to let the mother apply the mental tests, lest the child be unduly disturbed by the stranger.

Put a colored object in baby's hand, then place in front of it some article of food baby is especially fond of—note if it drops the toy and reaches out for the food.

Let mother encourage her baby to clap hands, shake “bye bye” and perform similar “baby tricks”—note its power of imitation.

### *Mental Age, Eighteen Months*

Engage baby in simple games, such as throwing ball and the like—note its dexterity.

Hand baby a pencil and some article of food—note its understanding of their use.

Let mother encourage baby to repeat “papa,” “mama” or similar words—note its power of articulation of syllables and words.

### *Mental Age, Two Years*

Learn whether baby knows its own name and that of mother, brother or any other member of the family.

Hand baby some article of food—note its power to bite and to masticate.

Put in front of baby some constructive toy—note its power to manipulate the same, e.g., to “build a house” of wooden blocks.

Ask baby to point to his nose, mouth, eyes, etc.—note response.

*Mental Age, Three Years*

Encourage baby to repeat several numbers or short nursery rhymes it was taught to recite, or to sing—note his power to memorize.

Place child in front of window and let him tell you what he sees on the street—note his ability to distinguish men from animals or objects.

Show him a picture-book with different animals and ask him to point to a horse, cat, bird, etc.—note the ease of response.

Display several pictures of relatives and let him pick those of parents.

Direct him to bring you different small objects from bureau drawers or closets—note his way of going about it and the ease with which he locates them.

If already instructed, ask him to spell his name, to count, etc.—note his memorizing power.

*Mental Age, Four Years*

Test his ability to feed himself with spoon or fork.

Let him reply to the following questions:

Where do you live? Where do you sleep? What did you have for luncheon to-day, or yesterday? How old are you?—almost all normal children of four years and younger are able to promptly respond to these questions, or to similar ones.

Let him pick out several letters of the alphabet, especially those required to spell his name—note the ease with which he accomplishes it.

In addition to these simple tests which serve to establish the diagnosis of amentia in general, we have a number of pathognomonic clinical syndromes which in view of their usual occurrence with certain lesions in the brain and ductless glands, enable us to classify amentia in the following distinct clinical groups, viz.: Amentia symptomatic of microcephalus, hydrocephalus, and cerebral hemorrhage and inflammation; amaurotic family idiocy; Mongolism; Cretinism and myxidiocy; infantilism, and mental retardation from other causes, all of which will presently receive full consideration.

## CHAPTER III

Special Groups of Amentia. Microcephalus. Hydrocephalus. Paralytic Amentia. Amaurotic Family Idiocy. Mongolism. Cretinism. Infantilism. Moramentia. Their Differential Diagnosis.

### MICROCEPHALUS

FROM a fairly large number of cases under my observation I have been tempted to distinguish two forms of microcephalus. One, in which the brain as a whole is very miniature, but not deficient in its component parts, thus showing arrest of development, but not a state of disease. The second variety is characterized by an absence or degeneration of several components of the brain, such as the peduncles, pyramids or even an entire hemisphere. In these cases there may even be an hydrocephalus in conjunction with the microcephalus. In the first variety of microcephalus (see Fig. 2) the skull is thick, very small and sometimes deeply furrowed. The cranial sutures are effaced and the fontanelles completely ossified. In the second variety (see

Fig. 3), the reverse may be the case. Indeed, in some microcephiles the skull may be moderately large and irregular in shape ("dome" or "sugar-loaf"). In microcephiles there is often also a hypoplasia of the spinal cord, more especially of the pyramidal tracts and the columns of Goll.

Where the brain is intact but miniature there is general inactivity of the cerebro-spinal system. The child is entirely helpless during infancy, but occasionally gradually improves physically as he gets older. As the cranial bones are completely ossified, and the immature brain no longer has the facility to develop, the mental faculties of the child remain permanently in an infantile state. On the other hand, in the second variety of microcephalus the mental and physical condition of the child depends entirely upon the pathologic alterations of the brain. Where the motor area is involved, we have disturbances of locomotion, convulsions athetosis, rigidity and many other symptoms that usually accompany cerebral lesions. The mental state of the child ranges from feeble-mindedness to profound idiocy. As these children get older, they are usually obstinate, vulgar and very irritable. Some of them understand simple words addressed to

them and are able to imitate certain actions after prolonged training. They may learn to feed themselves, to do little errands, and possibly to help in some trade under the guidance of a master. The majority of them, however, are entirely devoid of understanding and take no interest in their surroundings, and especially while under three or four years of age, may for hours sit or lie in one position and indulge in irregular movements, without by attitude or facial expression indicating any desire for a change or even betraying any discomfort during or after defecation or urination. Owing to their extreme restlessness and awkwardness of locomotion, their grotesque movements in hopping from place to place often resemble those of rabbits, goats or monkeys, and in times bygone were exhibited by showmen as curious descendants of a lost, degenerated race. Some of them are witty and alert and show distinct powers of mimicry, but they never attain a sufficiently high degree of intelligence to independently earn a livelihood.

The diagnosis of microcephalus is based principally upon the size and shape of the head. In the first variety of microcephalus, where owing to early arrest of development of the brain the

cranial bones ossify before or immediately after birth, the circumference of the skull always remains from 3 to 6 inches below that of the average normal child. To a slighter extent (2 to 3 inches) this is true also of the second variety of microcephalus. The hair is often so coarse and wiry that, as Tredgold puts it, the teeth of the clippers often break whilst the hair is being cut. A microcephalic idiot may sometimes be mistaken for a Mongolian. In Mongolism, however, the head is not quite as small or malformed, the hair not as coarse, and the muscular flaccidity or rigidity not quite as pronounced, while in microcephalus protrusion and cracking of the tongue is exceptional. In early infancy the mentality of the Mongolian is on a higher plane than that of the microcephalic idiot. The flaccid type of microcephalus may occasionally resemble amaurotic family idiocy. In the latter condition, however, there is usually a history of gradual degeneration after birth: the fontanelles are usually open, the size and shape of the head fairly normal, and an ophthalmoscopic examination reveals pathognomonic changes in the retina (see p. 79). The aforementioned symptoms of microcephalus are also ample to differentiate this form of amentia



from that associated with traumatic cerebral palsy. Besides, in Paralytic amentia of natal origin, congenital stigmata of degeneration (see p. 25), which almost invariably prevail in microcephalus, and the "idiotic grunt"—the guttural noise which the microcephile usually exhibits, particularly when he is enjoying a square meal—are usually absent. Finally, it is well to bear in mind that "sugar-loaf" head (oxycephaly) is occasionally met in perfectly normal children.

#### *Illustrative Cases*

Fig. 2. H. B., 4 years old, fifth child of Jewish parents, who seem to be in good health though there is a positive history of tuberculosis and neurasthenia in their immediate ancestry. Two of their children died supposedly from gastroenteritis in early infancy. This child weighed about six pounds at birth, and was apparently perfectly normal except for the small size of his head which was often commented upon by those who first saw him. When he came under my observation his head measured  $14\frac{1}{2}$  inches in circumference, but was free from asymmetry or malformation. He was able to sit up and stand when supported; he was docile, apathetic, and

took no interest in the surroundings except when accosted by his mother, when he seemed to recognize her voice and utter some sound apparently



FIG. 2.

of satisfaction. If left undisturbed, he would for hours lie or sit in the one position, play awkwardly with toys put in his hands. He showed no spontaneity whatever, except when he became very hungry, in which event he would be very restless and cry occasionally until his appetite was appeased. His appetite was poor, and he manifested no desire for any special articles of food. He had to be fed, principally on a soft diet

owing to his inability to bite and grind solid food. His teeth were very frail and irregularly implanted.

Fig. 3. J. B., 15 months old, third child of Russian-Jewish parents. There was a definite history in the mother of dreadful agony during

pregnancy. While four months pregnant a terrible Jew-baiting pogrom broke out in the Russian town she lived in, and not only was she herself badly beaten up and for days had not tasted a morsel of food, but was the eyewitness of the most cruel deeds perpetrated by drink-crazed, bloodthirsty mobs. She became a nervous wreck and for months thereafter was unable to obtain a few hours of undisturbed sleep. But notwithstanding all this the child was supposed to have weighed nine pounds at birth. It did well physically on the mother's breast,



FIG. 3.

but it seemed to grow more stupid from day to day. It was a very restless creature, for ever shaking and biting its hands and fingers, and

paying absolutely no attention to anything or anybody. His head was asymmetrical and dome-shaped and measured 16 inches in circumference. He had twelve teeth, most of them badly decayed and irregularly set. His appetite was voracious and his general health seemed to be perfect.

### HYDROCEPHALUS

The pathologic anatomy in hydrocephalic amentia varies greatly with the quantity of cerebrospinal fluid in the cranial cavity, the period of its appearance and the length of time it has continued to exert pressure upon the vital structures of the brain. Thus do we find that in cases of postnatal hydrocephalus, where the pressure happens to be slight and temporary, the pathologic alterations in the brain are often insignificant, whereas in marked congenital hydrocephalus postmortem examination usually reveals considerable atrophy of several parts of the brain. The brain markings are generally effaced, the ventricles distended and their contiguous structures compressed and degenerated. The meninges are thin and bulging, the cranial bones greatly atrophied, and the fontanelles and

sutures widely gaping with Wormian bodies freely distributed in the intervening spaces. Not rarely hydrocephalus is associated with spina bifida—undoubtedly nature's attempt to relieve the excessive intracranial pressure (see Fig. 4).

The most striking physical sign of hydrocephalic amentia is the extraordinary size and shape of the head. The head is usually asymmetrical, twisted in appearance (plagiocephalic); but may be rounded, egg-shaped (brachycephalic), long and narrow (dolichocephalic), or keel-shaped (scaphocephalic). The circumference of the head ranges between 22 and 30 inches or more. The scalp is very thin and barely covered by fine hair and traversed by conspicuous veins. The cranial bones are soft and often yield to light pressure with the finger, imparting the sensation of parchment. In severe cases the orbital plates are pushed downward while the eyeballs protrude forward, so that the lids are more or less retracted leaving a ring of the sclerotic exposed. This anomaly gives rise to the peculiar staring expression of the eyes which is characteristic of the hydrocephalic idiot, and is especially pronounced when accompanied by strabismus and nystagmus.

The mental symptoms are not invariably correlated to the size of the head, some infants with huge heads occasionally possessing more intelligence than those with proportionately smaller heads. And if, perchance, the hydrocephalus is arrested before permanent damage to the brain has been wrought, the hydrocephalic may yet grow up with a fair degree of mental capacity. Ordinarily hydrocephalic infants are quiet, gentle, timid, sorrowful and affectionate, and but little impressionable or curious. Owing to impaired function of the extremities by paraplegia and spasmodic contractures of the arms, they are rarely able to walk about and to help themselves, and when, as is often the case, vision (optic atrophy) and hearing are affected, they usually remain infantile for life—which latter, fortunately, is very rarely of long duration. Occasionally hydrocephalus is associated with adipsitas (see Fig. 5), which most probably occurs in consequence of interference with the functional activity of the hypophysis cerebri.

Chronic hydrocephalus may be confounded with rachitis, syphilis and macrocephalus in connection with hypertrophy of the brain. In

rachitis the extremities are weak, but neither paralyzed nor rigid, while mental deficiency, if present, is but slight, in hydrocephalic amentia the reverse is the case. The rickety head never attains the size of that of the hydrocephalic, and the cranial bones rapidly assume their normal consistency upon removal of the cause of rickets, i.e., on attention to hygiene and proper nutrition and administration of lime and phosphorus. In rachitis we usually find that in the first few months of its existence the child's physical and mental condition was normal, whereas in hydrocephalus there is a history of the presence of all of the aforementioned symptoms from birth on, or their sudden development in connection with some serious acute affection, especially tuberculous or cerebrospinal meningitis. Moreover, in these cases the child rarely escapes severe involvement of the eyes and ears. In syphilis the head is not rarely greatly enlarged, but instead of being unusually soft it is often hard and bossed. Of course, where the syphilitic is also suffering from hydrocephalus—which is not at all uncommon (see Fig. 7)—the differential diagnosis between these two forms of amentia can be made only by means of Wassermann's



reaction, which should at any rate be employed from a therapeutic point of view.

The following suggestions will prove helpful to differentiate hydrocephalus from macrocephalus associated with hypertrophy of the brain.

HYPERTROPHY OF THE BRAIN.	HYDROCEPHALUS.
The cranial bones are usually normal in consistency; the enlargement develops slowly.	Usually the reverse.
Marked pulsation at the anterior fontanelle.	Slight, if any.
Sutures slightly disconnected.	Widely gaping.
Ordinarily normal mentality or only slight deficiency.	Idiocy, as a rule.
Slight pressure symptoms, if the fontanelles are open.	Very marked.

### *Illustrative Cases*

Figs. 4 and 5. M. C., 8 months old, the only child of Italian parents. The history revealed insanity on the father's side, and early death of the mother's parents, supposedly from febrile diseases. The child was well developed at birth except for the hydrocephalus and spina bifida. Wassermann's reaction was negative. The baby's mental condition varied greatly with that of the spinal hernia. Every few weeks the her-



nial sac would fill up with cerebrospinal fluid and rupture. A few days before its occurrence, the child would become very restless, refuse food, cry almost incessantly, and suffer from slight convulsive movements of the face and extremities. Immediately after rupture of the sac and



FIG. 1.

escape of a large quantity of cerebrospinal fluid, the child would brighten up and in almost every way act like a normal infant of the same age or thereabouts. At no time, however, was the child able to hold its head erect or to make free use of its extremities. The spinal muscles were especially weak, and when the support was taken off the head, the latter promptly fell backward

with a thump. The cranial circumference of the head ranged between  $19\frac{1}{2}$  and 22 inches, depending upon whether the measurement was taken



FIG. 5.

of Jewish parents. The baby was normal at birth and the family history presented nothing remarkable, except that the father never earned enough to support his ever-growing family. The child was breast-fed and seemed to do

before or after the escape of the cerebrospinal fluid. Were it possible to arrest the cause of the hydrocephalus, an operation for the cure of the spina bifida would have been indicated. In this event also it is quite probable that the child's mentality would have become normal, since the intracranial pressure had at no time reached such force, apparently, as to destroy cerebral structures.

Fig. 6. M. C., 15 months old, sixth child

very well, until about four months old, when he was suddenly seized with convulsions, high temperature and semicoma. The condition was diagnosed as meningitis, and after a stay



FIG. 6.

of six weeks in one of the city hospitals he was returned home, seemingly in fairly good health. The mother noticed that the head was unusually large and that the baby did not act quite right, but appreciating that he had just passed through so serious an illness, she was re-

signed to wait for gradual improvement. This, however, never materialized, for when it came under my observation, there could be no question but that we were dealing with a hydrocephalic idiot. The head measured 23 inches in circumference; the anterior fontanelle was widely open, and the cranial bones were unusually soft, giving the sensation of parchment paper to the palpat-ing finger. There was pronounced nystagmus, protrusion of the tongue, general muscular insufficiency and total indifference to his surroundings. He was free from any signs of rachitis and organic affections and weighed 24 pounds. He was happiest when left alone.

Fig. 7. F. K., 11 months old, second child of Irish-American parents. The first child died soon after delivery, and there is a history of a miscarriage previous to the birth of the first child. The baby was born at full term and appeared to be perfectly normal. It was breast fed and thrived for several weeks. It was soon noticed, however, that its head was growing exceptionally fast, and the baby was gradually losing its power to hold the head erect. As at the same time the child was also gaining in weight and length, slept well and was free from any appreciable

symptoms of ill health, the parents did not deem it necessary to consult a physician. As months passed by, they observed, however, that the baby's mental development failed to reveal itself, and that its body musculature remained extremely weak, and hence they consulted a physician who diagnosed the case as rachitis and recommended appropriate remedies for this condition, which of course, remained ineffectual. When it came under my observation the circumference of the head

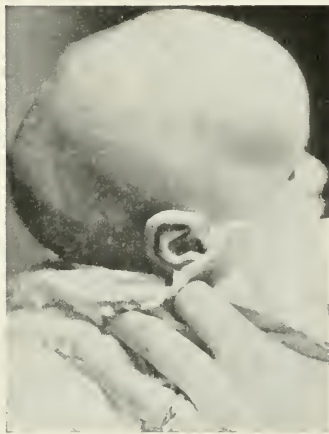


FIG. 7.

measured over 23 inches. The cranial bones were thin in some spots and markedly thickened in others: the anterior fontanelle was widely gaping and greatly distended. The covering skin was very thin and tense and traversed by dilated veins. The child was able to hear and to see and seemed to recognize the mother, both by the tone of her voice as well as when

she approached him. This was practically all he was able to do, and his mentality in general equalled that of a three-months-old baby. The diagnosis of syphilitic hydrocephalus appeared very plain, and a Wassermann test confirmed the diagnosis. Under the circumstances, therefore, I put the child on an anti-syphilitic course of treatment, consisting of inunctions of mercury and sodium iodid internally, but although the treatment was continued for six weeks, no appreciable improvement in his condition could be noted. On two occasions also I employed lumbar puncture followed by firm strapping of the head. But it took only a few days for the fluid to reaccumulate, and I was obliged soon to remove the adhesive straps, since twitching of the eyes and extremities set in, and the child was made very miserable. I next suggested the injection of neosalvarsan, but the parents thought that I had experimented on the baby long enough.

#### PARALYTIC AMENTIA

*(Vascular, Inflammatory, Toxic, Meningitic, Epileptic Amentia)*

Under this heading are generally grouped the numerous cases of mental deficiency which are

due to more or less extensive lesions in the brain occurring either before and during birth of the child or in the course of its first few years of life. The cerebral lesions may be the result of hemorrhage or inflammation, or both, accompanying prenatal or postnatal cranial traumatism, asphyxia neonatorum, meningitis, encephalitis, measles, scarlet or typhoid fever, pertussis and similar microbial affections, and neoplasms. In the great majority of these cases the cranial bones are reduced in thickness, the meninges are adherent, and some of the convolutions are compressed, atrophied and indurated. Some portions of the brain are in a state of softening, others are found to have undergone cystic degeneration, cicatricial contraction and sclerosis. The lesions are productive of variable clinical pictures in different individuals. They may lead to paralysis in one child, convulsions in another, and to amentia in the third or to all these manifestations in one and the same child. Moreover, these phenomena are not invariably correlated to the extent of the lesions. And one is occasionally surprised to find diffuse lobular sclerosis of the brain with extensive blood cysts and porencephaly in a child who during life was apparently en-



dowed with fairly normal mental faculties, and, conversely, only minute cerebral lesions with total idiocy. As a rule, however, in infants any seemingly trivial intracranial accident is followed by mental deficiency, hemiplegia or diplegia, and this is especially the case with lesions in the frontal, prefrontal and parietal lobes.

Of 55 cases of hemiplegia examined by Sachs and Peterson, the mental impairment was feeble-mindedness in 16 children, imbecility in 31, idiocy in 7 and epileptic insanity in 1 case. In diplegia the percentage of mental deficiency is always very high, between 60 and 75, whereas in cerebral paraplegia, which condition is usually associated with less extensive lesions, the mental deficiency is rarely very pronounced. Epilepsy, especially of the Jacksonian type, is quite a common sequel of cerebral hemorrhage or inflammation and ultimately ends up with progressive amentia.

In order to obtain a clearer conception of the symptom-complexes that usually follow the aforementioned pathologic alterations in the brain, it is advantageous to classify these cases in three large groups—in accord with the time of their development, either before, during or after birth. The first group usually reaches this



world in a more or less abnormal physical condition. They are often prematurely born, emaciated and disfigured, and of very low vitality. The head is either small and asymmetrical or normal in size, but soft and flattened on one side. The extremities are either rigid or slightly movable or there may be mono- para- or di-plegia. As these symptoms are the result either of arrested development of the brain or cord, or of both, or hemorrhagic or inflammatory processes therein, it is not at all uncommon to find several cranial nerves implicated. Under these conditions, of course, the diagnosis of congenital paralytic amentia is self-evident. The second group is generally described as Little's disease, or diplegia or paraplegia spastica infantilis. In the great majority of these cases there is a history of natal traumatism, asphyxia, convulsions immediately after birth and other signs of acute cerebral involvement. Rigidity of the limbs appears either very soon after birth or not until several weeks or months thereafter. The child usually lies motionless, with its legs either pressed against each other or one upon the other. As it grows older it is noticed that it does not attempt to stand or walk as early as a normal baby, and that

the feet seem to stick to the floor during its attempt to move its legs forward. If it gradually manages to walk, it takes short, rigid steps, with the feet in tiptoe position and the knees pressed closely together or the legs crossing each other (scissors-gait), sometimes half running, so that at every step falling seems imminent. The rigidity often gradually gets worse, extends to the extremities and trunk, if not already affected, and leads to fixed deformities. A Z-shaped deformity is often observed in the hands, when the child attempts to use them. Early in the course of the affection the deformities may disappear during sound sleep or under anæsthesia. The knee-jerk is exaggerated and ankle-clonus is often very pronounced, while muscular strophy is slight and the sphincters are but little affected. This characteristic physical syndrome is invariably associated with mental deterioration, ranging from simple feeble-mindedness to total idiocy, and is often accompanied by stammering, nystagmus, strabismus, athetosis and epileptic convulsions. The third group of cases gives a history of apparently normal physical and mental development at birth, and of an acute or insidious onset of some febrile or wasting disease or of

traumatism some time after birth, which were later followed by amentia with or without paralysis or epilepsy, and often degeneration of cranial nerves. The mental impairment is usually progressive in character, and in older children may not be fully recognized until several months or years after the accident or termination of the primary affection. To this group of cases, encephalitis and meningitis and their prolific sequelæ, more especially partial or total deaf-mutism and blindness, contribute the greatest number of victims, although traumatism with its great tendency toward epilepsy is exceedingly conspicuous in the histories of postnatal amentia recorded. According to Fletcher Beach, other infectious diseases, such as typhoid, scarlet fever and measles, do not form very rampant causes of this form of amentia, for after examining the histories of 2,000 cases of idiocy, imbecility and feeble-mindedness, he found only 37 (or 1.85 per cent.) which could be traced to an attack of one of these affections.

In connection with paralytic amentia it is opportune to call attention to a form of mental backwardness which is occasionally encountered as a result of hereditary syphilis. The amentia

may appear, as has already been stated, in consequence of hydrocephalus (see Fig. 7) or in connection with mono- hemi- or di-plegia consecutive to syphilitic meningitis or encephalitis. More rarely the foundation to the amentia is established during intrauterine life in the form of gummatous infiltration and sclerosis of the brain. In this event the child is born with all the symptoms corresponding with the primary lesion in the brain, e.g., paralysis, defective vision or hearing or involvement of other cranial nerves. Except for the history of the case and the positive Wassermann reaction there is practically no way of distinguishing syphilitic from non-syphilitic paralytic amentia. However, irregular enlargement of the head, particularly Parrot's nodes, should serve to arouse our suspicion.

#### *Illustrative Cases*

Fig. 8. S. P., 8 months old, the only child of Polish parents. When six months pregnant the mother was severely injured by a speeding automobile. She was laid up in bed for about two months, was threatened with miscarriage, but the baby was finally born about two weeks

before full term and weighed 6 pounds at birth. It may also be of interest to note that the mother was 38 years old, 12 years married before the child was born. The diplegia was present at birth, but outside of a slight deformity of the ears no stigmata of degeneration are discernible. The baby is totally idiotic, is unable to hold the head erect—in fact, the head is in a more or less constant state of opisthotonos—and is subject to several attacks of convulsions every few days. It is extremely sensitive to sudden noise or manipulation, both of which are not rarely followed by convulsive seizures. Ophthalmoscopic examination failed to disclose any abnormality of the retina or optic nerve, but this notwithstanding,



FIG. 8.

we have been unable to attract its attention even momentarily. It manifests its desire to feed by extreme restlessness.

Fig. 9. Z. R., 30 months old. She was normal at birth, weighed over 9 pounds. The family history was negative, except for the fact that father and mother, both Irish-Americans, were



FIG. 9.

habitually partakers of fairly large quantities of liquor, and that the father on rare occasions drank to excess. The baby was breast-fed up to five months of age, and received a mixed diet thereafter. She had several teeth, crept, and

walked with little support before she was one year old. When about 20 months old she was seized with an attack of influenza which was apparently complicated by encephalitis, for it was followed by paralysis of the left abducens and facial nerves and very slow convalescence. During this period it was noticed that she was completely changed in her behavior as compared with that previous to the attack. She was greatly depressed and irritable and cried without

provocation. At first this demeanor was attributed to weakness, but it was soon found that although gradually gaining in physical strength, her mental faculties have utterly failed to keep pace with her physical progress, nay, seemed to be retrogressing from month to month. When she came under my observation her mental age equaled that of a one-year-old child; her speech was very defective—partly probably due to the facial paralysis—she was very slow in responding to simple questions, and showed no inclination to speak, though before taking sick the contrary was the case. There was distinct paresis of the right side of the body, which fact, together with the left facial and abducens paralysis, indicated that the lesion was in the pons and medulla. After several months' treatment with massage, electricity, hydrotherapy and nerve tonics, there was considerable improvement in the muscular activity of the child, her mentality, however, remained stationary.

Fig. 10. J. B., 5 years old, the only child of German-American parents. His father died from hydrophobia. The mother is very delicate in health and suffering from chronic mitral regurgitation. The boy was delivered without



instruments and seemed normal physically, but weighed only six pounds. The child was always puny and began to walk at about two years of



FIG. 10.

age, when the mother noticed that his legs were rigid, that he could not walk unless supported, and even then crossed one leg over the other and dragged them along the floor. On examination I found marked spasticity of the lower extrem-



ities, greatly exaggerated reflexes and ankle-clonus. He has an awkward, hobbling gait, tires easily, and often falls forward. His upper extremities also are impeded in their action, and his hands often assume a Z-shaped appearance, when strenuous voluntary motion is attempted. His mentality equals in acuity that of a three-year-old child. It is difficult to tell, however, how much of the deficiency is due to faulty environment. Namely, the mother works out by the day, and the child is entrusted to the care of kind neighbors who are neither capable nor inclined to give the boy suitable training or education. Believing the case to be amenable to surgical treatment (decompression, tenotomy, etc.), I suggested to put the child in the hospital for operation.

Fig. 11. M. F., 27 months, the second child of Irish parents. The first child died a few days after birth. Father and mother were both heavy drinkers and led a miserable life. The little girl was delivered with instruments and weighed only four pounds at birth. Convulsions set in immediately after birth and continued to recur about every four weeks. Her eyes became infected by gonorrhea during delivery, leaving two large

staphylomas behind which greatly interfered with vision. There was also ptosis of the right lid, which was due either to the gonorrhoeal ophthalmia or to injury by the instruments. She was able to make free use of her extremities,



FIG. 11.

although she was very weak on her legs and unable to walk about without support. Her mentality was very backward, barely equaling that of a one-year-old child. She was able, however, to repeat a few short words, and had neither protrusion of the tongue, abnormality of the hair, nor the idiotic grunt. Rarely free from an eczematous eruption, the latter seemed to serve as the por-

tal of entry to the tubercle bacillus. A tuberculous lesion appeared at the left angle of the mouth and within six weeks she succumbed to general miliary tuberculosis. Judging by the total absence of paralysis of the extremities, I am inclined to believe that the amentia in this case was due to prenatal rather than

natal or postnatal influences, although it is quite probable that the trauma during birth and the miserable environment in which the infant lived acted as aggravating factors.

#### AMAUROTIC FAMILY IDIOCY \*

This form of amentia is based upon specific pathologic alterations in the brain. It is characterized by some degeneration in the cerebral white fibres throughout the course of the pyramidal tracts, in the inner capsule, crusta, pons and medulla, and also of the pyramidal tracts in the lateral as well as the anterior columns of the cord. Furthermore, the same changes are found also in the gray matter of the central nerve system—in the cortex of the brain, in the cranial nerve nuclei, and in the gray matter of the spinal cord down to the lowest lumbar and sacral segments. Wm. A. Holden has further established the fact that the changes in the retina are identical with those in the brain and cord, and were due to a degeneration of the retinal ganglion cells. Hirsch, after a very ex-

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\* Warren Tay described this affection in 1881 as a purely local inner eye disease, while B. Sachs, in 1887, recognized and described it as a distinct brain affection.

haustive histologic examination of several cases under his care, concluded that not only are the cells of the cortex of the brain affected, but the ganglion cells of the entire nerve system, the main features being a condition of chromatolysis and other degenerative processes of protoplasm, combined with considerable swelling of the cell-body and displacement of the nucleus toward the periphery of the cell. The neuroglia and the blood-vessels are found to be perfectly normal.

Like the pathology, the physical and mental characteristics are entirely pathognomonic. The apparently normally born and developing infant begins to fail in strength as it reaches the age of six or eight months. Although not losing in weight, nay, sometimes even gaining, it is noticed that the baby is unable to hold up its head, to sit erect, firmly to grasp objects placed in its hands, and even forcibly to suck on the nipple of breast or bottle. Simultaneously with the muscular atony the baby begins to lose interest in its surroundings, fails to smile when accosted, and to follow bright objects to which its attention is being directed—all indicating mental deterioration. When the backgrounds of the eyes are examined, a very peculiar retinal image is

obtained. Namely, the maculæ are cherry red in color and surrounded by large grayish-white patches. The optic nerves are atrophied in the great majority of cases, and there is often also strabismus and nystagmus. These eye-symptoms gradually lead to total blindness, and the muscular atony rapidly borders on paralysis. Hearing at first is hyperacute, but in the later stages of the affection becomes obtuse. At this time, also, there is often inordinate "explosive laughter," difficult deglutition, and a marked tendency to recurrent convulsions. In one case I noted pronounced hirsuties over the greater portion of the body. Thus deprived of sight and partially of the sense of hearing, limp and languid as a result of the ever-increasing atony of its musculature, the helpless creature gradually loses all its other senses and, fortunately, also its life. This usually occurs before the child attains two years of age. More recently Vogt has described a "juvenile" form of amaurotic family idiocy which begins to manifest itself at a later age and runs a more protracted course. Its identity with the "infantile" form of the disease, however, is not generally conceded.

As its term indicates, this form of amentia affects several members of the same family, or those who are closely related, and shows a very striking predilection for offspring of the Hebrew race, more especially of immigrants from Russia and Poland. This peculiar family predisposition seems to confirm the view held by Sachs and others that amaurotic family idiocy is due to a congenital arrest of development, although the "juvenile" form seems to point to a toxæmic nerve degeneration of postnatal origin.

In the early stages of the disease amaurotic idiocy may readily be mistaken for rachitis, but in this affection the pathognomonic amaurotic eye symptoms are absent and the mental deficiency, if there be any, is very slight. Furthermore, rachitis usually sets in more frequently in infants over one year of age and the muscular atony of the trunk and spinal muscles is never so pronounced as to produce dropping of the head backward. More difficulty may be experienced in differentiating amaurotic family idiocy from cerebral neoplasms, be they syphilitic, tuberculous or malignant. I recall a case of gliosarcoma of the pons affecting a one-year-

old infant which was under observation of several pediatricists and ophthalmologists of note and diagnosed as incipient amaurotic family idiocy, none of them even suspecting the presence of a cerebral tumor. While in the latter affection optic atrophy is a common symptom, there is never a cherry red discoloration of the macula. Furthermore, in tumor the muscular atony, paralysis and convulsions are most apt to be unilateral in the beginning, and to gradually become bilateral, while in the amaurotic all the symptoms are bilateral right from the start. In early infancy Mongolian and amaurotic idiocy have two cardinal symptoms in common, viz., protrusion of the tongue and general muscular atony, which may lead to errors in the diagnosis. In such cases an ophthalmoscopic examination is decisive. It will also be found that in amaurotic amentia the tongue protrudes but slightly and inconstantly, and is otherwise normal in appearance, the reverse being the case in Mongolism. Furthermore, in the latter condition the hair is wiry and the hands are usually spade-like and the mentality deficient from birth on. As has already been stated, in all cases of doubt an ophthalmoscopic exam-

ination should invariably be resorted to before arriving at a positive conclusion.

*Illustrative Case*

Fig. 12. S. F., 11 months, third child of Austrian-Hebrew parents. The family history was negative and the brother and sister of the



FIG. 12.

little patient are normal in development. The baby was normal at birth, was breast-fed, and grew nicely physically and, seemingly, also mentally up to six months of age. It then weighed over twelve pounds, was able to hold its head erect firmly, and sit up with ease when slightly supported. Soon after, however, the parents noticed that the child was slowly but surely losing ground. It grew pale, flabby, and less active. When put on the lap, the upper body would fall either forward or backward. It lost



interest in everything and everybody. It rarely smiled, and did so only after special inducements. Bright objects no longer attracted its attention, and as time passed on it rarely showed inclination even to feed. As usual, "teething" was primarily blamed for the extraordinary change, particularly since it manifested a tendency to protrude its tongue, and the saliva was freely flowing from its half-closed mouth. The family physician, noting that the child snored noisily, suggested the removal of adenoids—which was done without the slightest improvement in that direction. Another physician thought that the baby was suffering from rachitis and suggested a liberal mixed diet. When it came under my observation it was nine and a half months old. The aforementioned clinical picture was then more typically expressed, and an ophthalmoscopic examination disclosed the pathognomonic cherry red spot at the macula with the surrounding grayish-white discoloration. As the father of the baby once served in the Austrian army and was very fond of "life," I thought it advisable to give the baby the benefit of a course of antisyphilitic treatment, but it proved of no avail. From week to week the

clinical features, especially the amaurosis and muscular atony and idiocy, progressively became worse. The patient finally contracted the grip, which rapidly led to hypostatic pneumonia and terminated fatally within a few hours.

#### MONGOLISM

Except for the proportionately undue smallness of the pons, medulla, and cerebellum in relation to the cerebrum as compared with those of normal babies, the central nerve system of the Mongolian idiot shows no characteristic lesions. As in other forms of amentia, the brain is immature and its cells are imperfectly developed. With growth of the body as a whole the brain, too, attains a higher state of perfection, but never is capable to unfold the faculties of normal intellect. Mongolism is frequently associated with anomalies of the thyroid gland (hence were Mongolians often described as "cretinoids") and of the heart, and not rarely with general tuberculosis. There seems to be an etiologic relationship between Mongolism and syphilitic heredity. Sutherland, for example, having found a history of syphilis in 11 out of 25 cases of Mongolism under his observation.

This form of amentia was first described by J. L. Down in 1866, calling particular attention to the facial resemblance of the members of this group of idiocy to those of the Mongolian race, such as the Calmucks and Malays. The typical Mongolian idiot has a small, egg-shaped (brachycephalic) head, covered by thick, wiry hair; small saddle-shaped nose which is bound laterally toward the eyes by distinct vertical folds (epicanthus); triangular nostrils; almond-shaped, often prominent eyes, with speckled irides and eczematous eyelids; flat, usually flushed face, with high cheek-bones; distorted ears; high, narrow palate; cracked, ever-protruding tongue, with markedly enlarged papillæ, and flabby and clumsy, spade-like hands and feet. In addition to these characteristics, the Mongolian idiot generally presents marked laxity of the articulations, so that the tips of the fingers may be hyperextended almost to touch the dorsi of the hands, and the feet may be brought up to the neck while he is in a sitting posture. The little fingers are usually very short and curved inward; the skin is dry, rough and hairy, and, owing to circulatory disturbances, the Mongolian idiot often suffers from chilblains and

cracked lips. He is seldom free from hypertrophied adenoids and its sequelæ, i.e., nasopharyngitis, bronchitis, or even recurrent pneumonia. The latter may possibly be also due to the frequently accompanying rachitis, more especially chicken breast, large abdomen and spinal curvatures.

Notwithstanding all their troubles, Mongolian idiots are of a happy disposition, placid and affectionate. For this reason parents often fail to recognize the abnormal state of their children, even though they note their general bodily weakness, more particularly their inability to sit erect, to stand and walk. At about two years of age these aments usually become more active, vivacious, mischievous, full of grimaces and facial contortions—often misleading the parents to believe that they had outgrown their tardy development, and even to assume that their children were exceptionally bright. However, as time goes on it is generally found that their mentality is practically at a standstill, that they rarely can understand when spoken to, and still more seldom are able to speak. It is not uncommon to meet with Mongolian idiots two or three years old barely able to repeat single syllables,

to feed themselves even with the fingers, or to respond to nature's calls. As they get older they learn to walk and to make themselves understood, and after suitable training to make themselves useful and to perform little acts for their personal comfort, but they always remain in a primitive mental as well as physical state of development: unreasonable, helpless, awkward and uncleanly, often acquiring vicious habits (e.g., masturbation), which help to undermine their frail constitutions.

With this clinical picture in view, there ought to be no difficulty to distinguish typical Mongolism from similar forms of amentia. Atypical cases, however, may be mistaken for cretinism, microcephalus and rachitis.

In microcephalus the idiocy is more pronounced, the head either very small or asymmetrical, and the ability to make free use of the extremities in grasping, standing and walking appears at a very much later age than in the Mongolian idiot.

Mongolism differs from cretinism in the following particulars:

## MONGOLISM.

Skull brachycephalic.  
 Hair straight, wiry and abundant.  
 Skin thin, hairy and mottled.  
 Face flushed, vivacious.  
 Eyes almond-shaped; epicanthus.  
 Tongue narrow, cracked.  
 Little finger curved inward.  
 Thyroid treatment futile, as a rule.

## CRETINISM.

Quite normal.  
 Fine and sparse.  
 Swollen, "padded."  
 Pale and apathetic.  
 Palpebral fissures  
 horizontal.  
 Broad, swollen, pale.  
 Stumpy.  
 Very beneficial.

We can readily distinguish rachitis from Mongolism by the fact that in this affection the head is more or less square, soft and covered by fine hair, more especially along the occiput; the eyes are normal, the face is pale, the tongue not protruding nor cracked, and the fingers are normal in shape. Rachitis may delay the cerebral functions for a few months but the powers of speech, perception and voluntary motion are intact, and the mentality of the rachitic child rapidly improves with the amelioration of its physical condition.

Finally, let me emphasize that a diagnosis of Mongolism should not be based upon the infant's physiognomy alone, for occasionally we may be confronted by a baby of Mongolian ancestry who may be otherwise perfectly normal in body and mind.

*Illustrative Case*

Fig. 13. R. G., 23 months old, fifth child of Jewish parents. There was nothing worth noting in the family except that the mother suffered greatly during pregnancy, was unusually large as to give rise to the suspicion of twin



FIG. 13.

pregnancy and was supposed to have had an excessive amount of liquor amnii. The baby weighed about 7 pounds, was breast-fed and seemed to thrive during the first few months. The parents noted the peculiar facial features of the baby early and wondered why the child persistently protruded his tongue, but accepting the view of their friends and neighbors that the child was "teething," they let, as they thought, well enough alone. Gradually, however, their



equanimity became greatly disturbed as they watched the infant sitting or lying in one position without showing the slightest interest in its surroundings and that at 15 months of age it was yet unable to say papa or mama. After consulting several physicians and carrying out different modes of treatment ranging from mercury—and iodides—saturation to thyroid feeding, it finally came under our observation. He looked the typical Calmuck, except for the wrinkled appearance of his forehead which somewhat suggested thyroid insufficiency. But after trying thyroid gland substance for six weeks without benefit and taking in consideration his wiry hair, cracked tongue, almond shaped eyes, extreme restlessness, the absence of fatty tumors, inward curving of the little fingers and marked separation of the large toes from the other toes, there seemed no doubt but that we were dealing with a case of Mongolian idiocy. The child presented also several symptoms of rachitis, such as large abdomen and enlarged epiphyses, but these manifestations could readily be excluded as the cause of the profound mental deficiency.



## CRETINISM—MYXIDIOCY

Thyroid insufficiency, though primarily not a brain affection, sooner or later gives rise to degeneration of the central nerve system, more especially of the cortical cells. In congenital cases the cerebrum is usually considerably smaller than in normal children, and its convolutions are simplified; the cerebellum is asymmetrical and its laminae are reduced in number. It is not rarely associated with hyperplasia of the pineal gland, the hypophysis and thymus gland, showing nature's attempt to compensate the thyroid insufficiency by hyperactivity of similar structures. Pathologically we distinguish two forms of thyroid insufficiency which lead to amentia.

1. Athyreosis or absence of the thyroid gland which is generally a congenital anomaly, but may exceptionally occur as a result of traumatism or accidental extirpation (*cachexia thyreoopriva*). In congenital athyreosis the gland is frequently found replaced by cysts or other neoplasms. Occasionally degenerated (or healthy!) thyroid tissue is implanted in the base of the tongue.
2. Hypothyreosis or deficiency of thyroid gland, which may be of antenatal origin (e.g.

congenital goiter) or develop later as a result of disease or traumatism. To this group belongs also the endemic form of goitrous degeneration of the thyroid which prevails especially in certain sections of Switzerland, Germany, Asia, England, Russia, Hungary and America, in shut-up valleys of mountainous districts, and is supposed to be due to some toxic substances in the unboiled drinking water.\* That goiter is not uncommon in very young infants can be gathered from the statistics collected by Denme who among 643 cases found 53 to be of prenatal origin, 37 which developed in infants under one month of age, 59 between two and twelve months and 35 between thirteen and forty-eight months.

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\* This view has recently been disputed and a number of clinicians look upon endemic cretinism as an infectious disease. In this connection the report of A. Kutschera (Wien. Klin. Woch. No. 45, 1910) is of considerable interest. He relates that he found two dogs to develop cretinism who shared the bed of their mistress, a senieretin. One dog was completely idiotic, could not bark and reacted to nothing. It had dry, brittle, dirty hair, and milk-teeth together with permanent teeth. After removing these two animals the author put in the cretin's bed a healthy four-months-old pup of healthy parents. After three months this pup developed a large head, and ten months later it became a full fledged cretin while the rest of the litter of the same parents who were not exposed to cretinic infection remained perfectly normal. A second animal of a large race which could not conveniently occupy the same bed with the cretin also developed normally. The author therefore believes that cretinism is transmissible by direct, close contact.

Postmortem examination discloses in cretinism marked alterations in the osseous system. The cranial bones are thickened, the diploë is diminished and according to Virchow the sphenobasilar suture prematurely closed. The long bones are thick and short and often markedly deformed. As in other forms of profound imbecility there is in cretinism retarded development of the centers of ossification of the carpals and of the epiphyses of the metacarpals and phalanges. Section of the tubular bones usually shows an invasion of fibrous tissue from the periosteum in between the epiphyses and shaft, thus hindering the growth of the bones in length. Around the base of the epiphysis there is sometimes a sheath-like prolongation which may even be ossified and form a distinct cup around the epiphysis. But in contrast to what is observed in rachitis, there is no proliferation of cartilage cells near the line of ossification. The same overlapping or cupping of the epiphyseal cartilages is noted also in the ribs and innominate bones and in the scapulæ.

The physical and mental manifestations of cretinism vary greatly with the degree of thyroid insufficiency. Moreover they set in at a later

period in breast-fed than in artificially fed infants owing to the fact that during the first few weeks of life breast-fed infants receive an ample supply of thyroid gland substance through the mother's milk to counteract their thyroid insufficiency. In acquired athyreoisism the characteristic symptoms of cretinism usually appear gradually, but once the clinical syndrome is completed, it is practically alike in the prenatal as well as postnatal cases. The head of the cretin is either normal in size or slightly enlarged, flat and plump and set upon a thick, short neck. The fontanelles usually remain open, the forehead is low, and the root of the nose is broad and sunken. The face is weak and senile. The eyelids and lips are edematous and the tongue is large and "swollen," and hence ever protrudes from the half-closed mouth. The teeth are slow in coming and rapid in decaying. The abdomen is greatly distended, often marked by a large umbilical hernia. The extremities are more or less deformed and the articulations thickened. The hands and feet are short and flabby. Cretins learn to walk late, and their gait is awkward and draggy. The skin is dry, waxy and doughy in consistency, and the hair is sparse and brittle.

The body temperature is generally subnormal, and owing also to the ever present anemia, cretins are very sensitive to cold, notwithstanding their corpulent appearance. "Fatty tumors" are usually found in the supraclavicular and axillary spaces.

The intelligence of the infantile cretin, as has already been stated, varies with the functioning capacity of the thyroid gland. In congenital athyreosis there is total idiocy (myxidiocy). Some cretins, the so-called semi-cretins, possess a fair measure of intelligence. They appreciate their surroundings, and are able to acquire a meagre vocabulary which may be ample to make their urgent wants understood, or even to reply to simple questions. On the other hand, where the thyroid insufficiency is marked, they never reach even this low state of mental development, and, on the contrary, get more stupid as they grow older. In the great majority of cretins, the special senses are implicated. Taste and smell are obtuse; hearing is defective, and vision dull. The voice of the cretin is ordinarily husky. Like hydrocephalic aments they are timid, gentle and unassuming, and if left untreated, they retain their childish behavior for life.

One of the most characteristic features of cretinism is its marvelous improvement under thyroid feeding. After exhibiting thyroid gland extract in one form or another for but a short time, the cretin is often transformed from an uncouth, apathetic and clumsy little creature into a lusty, gracile and growing human being. The blurred facial features gain youthful expression; the lustreless, withered hair takes on new life; the stunted stature shoots up to almost normal proportions, and the brutal stupidity gradually gives way to human intelligence. However, this marvelous transformation lasts only as long as the thyroid medication is permitted to exert its wonderful influence. With discontinuance of the treatment the cretin slowly but surely sinks back into his everlasting idiotic condition.

Total atyreosis in the early stages and partial cretinism at any period of early childhood may be confounded with severe forms of rachitis and Mongolism. The differentiation of the latter form of amentia from cretinism has already been discussed in connection with the former affection (see p. 88). In distinguishing cretinism from rachitis it is well to bear in mind that the latter may complicate the former disease. But

in rickets the deficiency of intellect is slight and not progressive, the tongue is neither large nor protruding; the skin is soft and thin and not rough and edematous; the hair is normal and bald only in spots, especially over the occiput, whereas in cretinism the hair is brittle all over the scalp, and finally the rachitic baby learns to talk early and its voice is perfectly normal even though it may be weak. Rachitis, complicated by congenital macroglossia and adenoids, may on very rare occasions lead to errors in the diagnosis, but careful inquiry into the history of the case and the exhibition of thyroid will soon clear up all doubts. Furthermore, it will generally be found that in congenital macroglossia the tongue gets gradually relatively smaller as the child's mouth grows larger, which is not the case in cretinism. Besides, there is always the marked difference in the physical and mental development of these children.

#### *Illustrative Cases*

Fig. 14. F. C., 6 months old, the second child of Jewish parents. The first child as well as the parents are in good health. The baby seemed normal at birth and took the breast well.



A few weeks later, however, the child's appearance began to change. She was getting unusually plump, though partaking of but little nourishment and occasionally suffering from indigestion.



FIG. 11.

She manifested a tendency persistently to protrude the tongue, to snore while asleep and to grunt while awake. The mother and good neighbors found plausible reasons for the plumpness and protrusion of the tongue, attributing the former to her unusually good temper (for she was forever sleeping), and the latter either to teething or, as the mother put it, to a "craving for something I must have

longed for during pregnancy—and could not obtain," but they could not explain the noisy breathing, and hence decided to call on a physician. After going the rounds from one clinic to another the baby finally came under my observation. It was a most typical example of myxidiosis. The face particularly was highly characteristic. It



was waxy, apathetic, lifeless, marbled, as it were, and much older in appearance for the child's age. The tongue was exceptionally large, pale and thick and snugly embraced by the deathly pale, unusually puffed lips. The posterior portion of the neck was short and thick, while the anterior portion, in the region of the larynx and trachea, was hollow, and the covering skin was wrinkled—apparently owing to total absence of thyroid tissue. The supra- and subclavicular spaces were filled out by the so-called fatty tumors. The abdomen was pendulous and flabby and marked by a large umbilical hernia. The hands greatly resembled the "trident hand," frequently encountered in achondroplasia. The mentality of the baby was not as bad as would ordinarily be expected with so typical physical manifestation of cretinism. She recognized the mother and showed great annoyance by the approach of strangers. Possibly she still received a little thyroid substance through its mother's milk. The infant improved considerably under moderately large doses of thyroid gland extract; a few months later, however, she succumbed to an acute attack of gastro-enteritis.

Figs. 15, 16, 17. L. G. was perfectly nor-

mal physically as well as mentally up to one year of age (see Fig. 15). The family history presented nothing worth noting except that the father was a Christian and the mother a Jewess. She was the fourth child and was born at full term, weighing about eight pounds. Without any apparent reason she slowly began "to forget"



FIG. 15.

the numerous little baby-tricks she was accustomed to perform, became apathetic, drowsy and dull, lost the power of sitting firmly, to rise from the pillow when in recumbent posture and to manipulate the toys she used to play with. With all this she

was steadily growing fatter, a coincidence which puzzled the parents as well as their medical adviser. Tonics, baths, and strengthening food failed to improve her debile condition, but, on the contrary, from month to month she became more helpless and more stupid. All this happened while they were living in Germany, and although frequently examined by leading clinicians, I understand, no one ever sug-

gested thyroid insufficiency as the cause of the child's degeneracy, nor was thyroid treatment instituted to remedy the trouble. When I first saw the little girl, she was about eight years old (see Fig. 16). She weighed 30 pounds and measured but 34 inches in height. She was unable to walk alone and dragged her feet along the floor if led by both hands. Her hair was sparse and thin; her teeth fearfully decayed and her gums spongy. Her skin was thick, dry and waxy. The body was plump and pseudo-lipomatous masses filled the supra- and infra-clavicular spaces. The anterior fontanelle was still open and the articulations of the extremities were thickened. Her abdomen was large and pendulous, but free from an umbilical hernia. Her face was senile, her lips were thick, but there was no protrusion of the tongue. Her mental capacity was that of a one-year-old infant. She retained her childish tastes; with difficulty she could repeat but a few incoherent words, and grinned whenever ac-



FIG. 16.

costed. I put her on five grain doses of thyroid extract twice a day, and after about eight weeks' treatment she gained two inches in length, began to walk quite briskly and in every other way



FIG. 17.

showed wonderful improvement physically as well as mentally (see Fig. 17). The improvement lasted while she was taking the thyroid and there was almost an immediate retrogression in her facial features and general behavior upon discontinuance of the treatment.

### INFANTILISM

Under this heading are generally grouped several types of abnormal infants who never attain the physical and mental development of adults and who retain several characteristics of infants and young children throughout life. In physiognomy and stature infantilism is closely allied to cretinism, and in many instances directly dependent upon thyroid insufficiency. Similar clinical syndromes have more recently been ob-

served in connection with deficient functions of the thymus, adrenals, pancreas, and pituitary gland, and some observers claim that similar arrests of development occur as the result of systemic poisoning by the syphilitic germ and other micro-organisms. In accord therefore with the aforementioned etiologic factors infantilism may be classified in the following types:

Thyroid-infantilism,  
Thymus-infantilism,  
Hypophysis-infantilism,  
Heredosyphilis-infantilism,  
Dystrophic-infantilism,  
Cardiac-infantilism,  
Intestinal-infantilism,  
Malarial-infantilism,  
Pellagra-infantilism.

Two special types of thyroid-infantilism are generally encountered, viz., Typus Brissaud, which is characterized by fullness of the face, plumpness of the body and clumsy extremities (see Fig. 18), and typus Lorrains, whose stature is gracile and whose facial features are pleasant and comely (see Fig. 19).

The subject in question is as yet awaiting considerable elucidation. With the advance of our

knowledge of the normal and abnormal actions of the ductless glands we shall undoubtedly be able to classify infantilism in two large groups, thus: Genuine infantilism, embracing all cases in which mental deficiency predominates, and a second form of infantilism, which is being described as microsomia, nanosomia, ateleiosis, asthenia, achondroplasia and Herter's infantilism, in all of which physical arrest of development predominates. I may state, by the way, that contrary to what is frequently recorded in medical literature, Herter's infantilism is not associated with actual mental deficiency. To quote this author, "the intelligence of these patients was in every instance good, although the necessity of living very carefully and obeying the directions of the physician and nurse has tended to make these children somewhat introspective as regards their own ailments and to form the basis of what might with increasing consciousness develop in after life into a hypochondriacal condition."

Congenital infantilism, like congenital idiocy in general, is often associated with physical stigmata of degeneration. Atrophy of the genitalia is particularly common where the ductless glands are involved. The mentality of these children

is very variable and depends entirely upon the period of life at which their mental development has been arrested. As a rule, they are never totally idiotic, and the majority of them are able to help themselves, to walk about and to play and to understand a simple conversation. Speech is usually delayed, but with advancing age and proper training, they ordinarily learn to speak, as well as to count and to write, and to earn a modest livelihood.

*Illustrative Cases.*

Fig. 18. J. P., 6 years old, second child of American parents. Father died soon after the child's birth of pulmonary tuberculosis; mother is insane and older brother is stupid. She



FIG. 18.



was artificially fed; began to walk and to talk at three years of age. She measures 32 inches in length; circumference of head  $19\frac{1}{2}$  inches; circumference of chest  $18\frac{1}{2}$  inches. Her mentality



FIG. 19.

equals that of a two-year-old baby. Destructive tendency.

Fig. 19. S. B.,  $4\frac{1}{2}$  years old, fourth child of Jewish parents. History of the case could not be obtained with any degree of accuracy. She measures 32 inches in height; chest circumference 19 inches; cranial circumference 18 inches; weighs 28 pounds. She is quite comely and affectionate, runs about without difficulty, and is able to feed herself. She cannot talk, but after persistent

efforts manages to repeat a few short words. She shuns her comrades but amuses herself with dolls and other toys. One of her thumbs is absent and the other rudimentary. X-ray examination reveals arrest of development of the carpal bones. Thyroid gland extract failed to show improvement in her mental condition which equaled that of a two-year-old infant.



## RETARDED MENTALITY \*

Delayed mental development is quite frequently the result of the following causes: 1. Deprivation of special senses, e.g., sight and hearing; 2, chronic affections, such as heart disease, and other severe nutritional disturbances; 3, faulty environment and education, or isolation.

Sense deprivation as a cause of retarded mentality need not be complete. Mere errors of refraction, for example, by leaving the child ignorant of numerous objects outside of its field of vision, may be entirely sufficient to delay the unfolding of its mental faculties. Similarly do we find that an infant afflicted with adenoids which interfere with acute hearing and renders it listless and inattentive, at least temporarily fails to receive the outside impulses to the brain and hence remains mentally backward. These children, however, are not suffering from amentia in the true sense of the word. On the contrary experience teaches that just as soon as the re-

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\* For this group of cases Moramentia would be a very appropriate term; mora (æ) signifying delay, impediment, hindrance, and mentia being used to designate mentality.

tarding elements are removed, e.g. removal of the adenoids, and correction of the visual defects, the supposedly mentally deficient children rapidly reach a normal state of mental development. As a striking corroborative illustration of this view, let me quote a few lines from ex-president Roosevelt's autobiography. After relating his varied experience as a mere boy, he emphasizes the point that quite unknown to himself he was under a hopeless disadvantage in studying nature. "I was very near-sighted, so that the only things I could study were those I ran against or stumbled over. . . . It puzzled me to find that my companions seemed to see things to shoot at which I could not see at all. One day they read aloud an advertisement in huge letters on a distant billboard, and I then realized that something was the matter, for not only was I unable to read the sign but I could not even see the letters. I spoke of this to my father, and soon afterward got my first pair of spectacles, which literally opened an entirely new world to me. I had no idea how beautiful the world was until I got those spectacles. I had been clumsy and an awkward little boy—a good deal of it was due to the fact that I could

not see, and yet was wholly ignorant that I was not seeing. The recollection of this experience gives me a keen sympathy with those who are trying in our public schools and elsewhere to remove the physical causes of deficiency in children who are often unjustly blamed for being obstinate and unambitious, or mentally stupid." Far be it from me to aver that had he been permanently deprived of acuity of vision, Mr. Roosevelt, because of this alone would have remained a mediocre man, but there is justification for the assumption that he would have lacked some of his brilliant faculties had he permanently remained near-sighted.

Defective vision, particularly if congenital in nature (e.g., congenital cataract) or acquired soon after birth, forms a greater impediment to normal mental development than a similar defect in the sense of hearing. It is quite common to meet with very intelligent deaf-mutes who by means of lip-reading or dactylology even in early childhood are able to make themselves understood and to fully express their wants. Very recently two deaf-mutes, brother and sister, came under my observation, who for intelligence could pass muster as any normal children of the same

ages. Their parents were first cousins, and their father was 15 years older than the mother. The little girl was eight years and the brother two years old. They had another brother, who was able to hear and to speak, but died at the age of five years during an attack of pneumonia. It was a most pathetic sight to watch the two children by means of dactylology and lip-language to converse among themselves or with their mother, and it was astonishing how much information the mother was able to convey to the little baby. The older child was full of life and possessed powers of observation and imagination rarely to be met in perfectly normal children of her age. While examining her she was intently interested in everything I was doing, and as I was testing her hearing—hoping possibly to detect a trace of it intact—she concentrated her whole mind upon the test, and off and on gleefully announced to her mother that she was capable of hearing—poor child, she was carried away by her vivid imagination! And I shall never forget her literally shining face and the grateful, almost overflowing eyes, when, to please her, she was assured, that her condition was not hopeless, and that it was merely a matter

of time when she would learn both to hear and to speak.

The mental deficiency encountered in children suffering from some chronic organic affections or nutritional disturbances (e.g., rachitis), like that associated with the aforementioned deprivation of senses, is only relative in character. In those children the brain possesses every potentiality for normal growth and development, but remains in a state of passivity for want of prerequisite outside impressions. This is due to the fact, on the one hand, that sickly, depressed children are not at all inclined to bother with what is transpiring around them, and, on the other hand, parents justly refrain from burdening sickly children with any sort of training and education. That deficient or disturbed nutrition, *per se*, is not a potent factor in the production of amentia, can readily be proven by watching the acuity of perception of emaciated, so-called marasmic, babies. Nothing that bears a semblance of food or its container escapes their attention, and they show a wonderful dexterity in manipulating the nipple or bottle at a very early age.

Faulty environment and isolation, like depriva-

tion of senses, greatly retards mental elaboration owing to lack of cerebral impressions by outside influences. We can hardly expect of a young child to distinguish objects it never had a chance to see or to touch; and the unfortunate baby that happens to be cursed with a habitually intoxicated father, mother, or both, and daily sees before it smashed heads and smashed dishes, and hears profanity on the one hand and incoherent babble on the other, is certainly ill prepared to acquire the attributes of normal mentality, and to show affection, power and imagination, judgment and discrimination. Faulty environment and isolation is not invariably the sad lot of the children of the poor and the degenerate. I have met with many a baby of fashion under two years of age or older, who at first impressed me as being utterly idiotic and who had remained mentally backward for several months thereafter, because of their having been secluded in some remote corner of their nurseries or huddled away under the upholstered hood of their carriages, and thus were given no opportunity to exercise their musculature or brain matter. Only too often do we see infants of the rich entrusted to the care of some inexperienced, half-baked, "white-linen-

nurses," who consider it their greatest achievement to keep the baby's bowels regular and have not the slightest conception of the importance of early mental development. But as has previously been stated this group of mentally backward children of the poor as well as of the rich, when by nature are endowed with normal brains, with marvelous celerity they retrieve the dormant mental faculties if placed in desirable surroundings and given the benefit of sensible management.

#### *Illustrative Cases*

Fig. 20. J. S., 22 months old, second child of Jewish parents. Father in second stage of tuberculosis, mother apparently well. The baby was normal at birth and did well up to eight months of age, when he was suffering from acute gastroenteritis. He never fully recovered from the attack, but soon began to show signs of rachitis and became subject to recurrent attacks of rhinopharyngitis, bronchitis and bronchopneumonia. When he came under my observation he weighed only 19 pounds. His facial features resembled those of an idiot and his mentality was very low. His nasopharynx was filled with hy-



pertrophied adenoids, and he had to keep his mouth widely open in order to be able to breathe. He was unable to stand alone, and his



FIG. 20.

entire vocabulary consisted of a few barely intelligible words. Throughout the body we could readily discern small subcutaneous nodules which in some places (e.g., the right hand) were in a broken down state. Apparently they were tubercular in character, for the tuberculin reaction was positive and there were no other tubercular lesions of the body. We excluded

idiocy by the history of the case, the absence of stigmata of degeneration or abnormality of the head (it measured  $18\frac{1}{2}$  inches in circumference) and by the fact that it spoke a little and re-



sponded to the mental tests of a nine-months-old baby. On removal of the adenoids and institution of a suitable régime as to hygiene and diet he brightened up very rapidly, which confirmed our view that we were here dealing with a case of retarded mental and physical development.

Fig. 21. J. W., 3½ years old, first child of colored parents. Father deserted the mother soon after the child's birth and the mother was obliged to put the boy in a boarding-house and to work by the day to earn a livelihood for herself and child. The baby was fed on condensed milk, patent baby foods and similar barely life-sustaining food products, and hence it early became an easy prey to rachitis with all its complications and sequelæ. When the photograph was taken he was still unable to stand without support, and practically every long bone of his body was infracted and deformed. Thus, stunted in growth and distorted in shape and features, a mere



FIG. 21.

glance at him told volumes of the misery he passed through in the few years of his existence. His mental age equaled that of a one-year-old child, except that the struggle for existence taught him to "beg" for food from all the boarders he came in contact with at the filthy lodging house, and to accost them in a language of his own which in time they seemed to have learned to understand. He was apathetic, listless, disinterested in his surroundings and apparently preferred to be left alone. But after being fondled for a while and spoken to in a friendly manner he could be aroused from his lethargy, to take notice of things shown to him and to manipulate little toys placed before him. His brain apparently was in a passive state because of lack of opportunity to develop, but capable of doing so in a suitable environment and with proper training.

Now let me relate one of the many experiences I have had with the poorly reared rich. It concerned the first child of wealthy, intelligent parents. The family history was negative. The little girl was born at full term and seemed normal in every way. The mother as well as the baby's nurse were very attentive to the child, but

lacked initiative and faithfully followed the directions of the family physician. The latter happened to be one of the medical automats frequently encountered in high society who in order to be up to date copy printed formulas when prescribing infant-foods and hastily adopt every fad and fancy of popular medical celebrities without individualization or discrimination. Laboratory milk having then been in fashion, the child, of course, was promptly put on this milk, its formula having been changed from time to time. When the baby came under my observation her dietary still consisted of this milk and a little orange juice. She was then 15 months old and weighed eighteen pounds. Her general musculature was very poorly developed; she was unable to sit erect without support, had no teeth and seemed entirely indifferent toward the parents or nurse. Toys barely attracted her attention and she manifested no desire to grasp food articles placed before her nor to put them in her mouth when brought near it. Her cranial bones were fully ossified and the circumference of the head measured 18 inches. The occiput was markedly flattened, but there were no signs of degeneration in any portion of the body. I learned that upon

the advice of the attending physician the baby was not allowed to sit up without support, "for fear of her getting a weak back," and to prevent the latter she was most of the time kept flat on her back, which fact was undoubtedly responsible for the flattening of the occiput. She was constantly sucking her thumb, a habit that she had acquired during the first few months of life. I understood that at that time the baby was frequently crying either from hunger or pain, and since the doctor had sternly forbidden the use of a "pacifier," explaining that this was the cause of adenoid vegetations (this is another fad that had met with popular credence, as though a clean, sterilized, if you will, nipple, was more harmful than a dirty thumb!), she found in the thumb a most convenient substitute. Now, seeing an infant with a flattened occiput, thumb in mouth, complete muscular atony, and above all, without the slightest indication of mental activity, I could not help but surmise some form of mental deficiency, probably of prenatal origin. Yet on careful examination no historical data nor physical signs could be discerned to substantiate such a presumption. After careful consideration, therefore, I came to the conclusion that

I most probably was dealing with one of those poorly reared rich children, and hence instituted a complete change in the bringing up of the child. In about three months' time the little girl was transformed from an inanimate, helpless and stupid little creature, into a vivacious, active and intelligent human being.

A striking example of the baneful effects of sense deprivation and faulty environment upon the mental development of the child has recently been recorded by Dr. Moreau of Saint-Etienne (*Loire médical*, abstr. *Jour. A. M. A.*, Oct. 25, 1913). The child was 8 years old when put in his service for operation on complete congenital bilateral cataract. Until entering the hospital the child had lived at home with his father, a poor peasant, and had received no training or education whatever. It was an anxious moment, when, a week after the operation, the bandage was removed from the eyes. The child on being able to see, made simply a few meaningless sounds. The bandages were replaced, and the trial repeated two days later, the child again showed no emotion. As each subject was shown to him after he touched it, so that he

knew it perfectly well, he said that he did not know what it was. It took many days for the child to acquire an idea of colors, but when he did, the color-concept assumed an abnormal significance, and everything was interpreted from this point of view. Whatever object was shown to him was white or black according to whether it faced the light or was in shadow. He analyzed everything that he saw into colored spots. Each new thing that he learned, however, was of use in appreciating things that came after; thus, when the child learned that the object was a box, everything empty that he could stick his hand into was for him a box; his drinking cup, his cap, his boots, etc. Whenever he wished to take hold of anything he acted as one does who can see, but in grasping it he relied more on the sense of touch than he did on sight. He had no sense of space. He tried to touch lamps which were a hundred feet away. He mistook the moon for a lamp and reached out for the stars. Fifteen months after his entrance into the hospital he could not read, in spite of the efforts of the nun who took care of him to teach him the alphabet. Then his father took him home. Dr. Moreau saw the child again a year later. He had remained all

this time without education, he had learned nothing new and, moreover, had forgotten most of the things he had picked up while at the hospital.

This case was certainly extraordinary. As a rule, when the avenues of transmission of impulses to the brain are opened children spontaneously acquire a certain amount of knowledge by imitation, even if no effort at all is being made to train or teach them. It is quite possible, however, that in this case the prolonged disuse of the brain cells gradually led to their atrophy or degeneration. That he was not suffering from genuine amentia we can gather from the facts that he was able to speak, and that during the time he was under the care of the nurse he readily responded to some outside impressions, which would leave a congenital idiot entirely unaffected.

## CHAPTER IV

Prophylaxis. Eugenics. Judicious Management of Pregnancy and Labor. Care of the Newly Born and of the Older Child. Mental Training of the Normal Infant.

THE aforementioned theoretical considerations of idiocy and the allied mental deficiencies in childhood tend greatly, I believe, to establish the facts, first, that amentia is preventable in a large proportion of cases, if prophylactic measures are instituted early; second, that under suitable management a great many mentally deficient children can be made useful to themselves and possibly also to the commonwealth. We shall now endeavor to offer a few practical suggestions to accomplish these highly desirable objects in view.

One of the most essential factors in the prevention of mental debility in the offspring is their inherent bodily and mental strength.

Inherent strength is not procurable after birth. It is a consummation, an inheritance of ancestral virility and vigor, premarital purity, conjugal devotion, matrimonial chastity, sobriety and ideal



hygiene. It can be fostered by sensible regulation of marriage, conservative mutual selection, avoidance of consanguineous mating and prohibition of marriage among those encumbered by chronic brain affections, grave wasting diseases, alcoholism, drug habits and extreme poverty. Above all, inherent strength can be fostered by judicious management of pregnancy, labor and the physical and mental care of the infant. Within recent years there has been a great awakening to the importance of exacting from those destined to procreate the race of the future that they be free from all encumbrances, congenital as well as acquired, which tend to embarrass their offspring in their normal development. And while eugenics as at present taught carries with it a good deal of useless, nay, harmful fiction and fetichism, which veil its true object and render it subject to ridicule and derision, there is every reason for the belief that after the noisy agitation has ceased and thorough sifting of the good from the bad has taken place, the world will be very much the better for it. In the meantime, or until the lustrous millennium has dawned upon us, it is entirely sufficient for physicians to preach practical rather than theo-

retical eugenics and to counsel those encumbered by grave hereditary taints to be very cautious in the selection of their mates, lest doubly marred heredity may intensify the degeneracy in the offspring. All agree that those suffering from specific venereal disease, tuberculosis, malignant disease, epilepsy and insanity are not marriageable subjects, and should not be permitted to marry unless they can show that they have remained free from any traces of these affections for a number of years. But it is not in the province of the physician to join eugenists in their hunt for "desirable types" of man- or womanhood, even were such types at all desirable. "What the eugenists set up as desirable types," says A. C. Jacobson, "strike many of us as merely smug, unctuously respectable and commonplace paragons. If the eugenists had their way and succeeded in peopling the world with a race of disgustingly normal beings, standardized to the Philistian scale which the intellectual plebeians who are so warmly drawn toward the eugenic camp seem determined to devise, life would be drab and jejune indeed. Happily such a consummation can never be, for which the gods be thanked. Anything approaching real control of

the race after the plan of these fanatical breeders is a phantasy . . . since haphazard 'scrub breeding' has gone on in the human family so long that pure strains with definite character units are practically unknown. Hence, who is the fit? What is fitness?" Physicians can do most good by judicious management of pregnancy and labor and the rearing of the child, more especially during its first few years of life. As has already been stated, after impregnation the destiny of the offspring is partially or wholly dependent upon the physical and mental welfare of the mother. "But even if it be proved—it has not yet been proved—that the conditions of life in the nine months before birth have no influence either for good or ill upon hereditary maladies and deformities, even then there remains much to be done in antenatal hygiene, for there cannot be the slightest doubt that many morbid influences come to play upon the body of the infant in the womb and that some at least of them may be prevented or their results cured" (Ballantyne). No definite statistics have thus far been adduced to show the degree or extent of the beneficial influence of antenatal hygiene upon the mentality of the offspring, but some approxi-

mate estimate can be obtained by analogy, when we compare the weight and physical power of resistance of babies born under favorable conditions with those born of mothers who up to the last moment of pregnancy were exposed to hardship and struggle for existence. Thus Pinard gives as the average weight of babies of women who worked up to the time of delivery about  $6\frac{1}{2}$  pounds, while for those born of women who had a short respite from hard work before delivery  $7\frac{1}{4}$  pounds. Bordé found the average weight of babies of Italian women who worked up to the delivery to be  $6\frac{1}{4}$  pounds, while of those who had rested a few weeks before delivery about 7 pounds. More recently S. Peller reported his findings among Austrian women. His material was drawn from two sources, a sanatorium for women of means with 612 patients, and a large clinic for poor women (under the direction of L. Teleky), with 4,875 cases. He found that the first born male babies of well-to-do women averaged about four ounces heavier and the female babies about three ounces heavier than the babies of poor women who worked up to confinement. Moreover, in a comparison of the first-born children of hospital women with those of

the women coming to the hospital just before confinement, the babies of the former are shown to average about four ounces heavier than those of the latter group. With these observations in view the importance of antenatal hygiene for the betterment of the race of the future becomes self evident. The prospective mother should be placed in a wholesome environment and proper hygienic surroundings. Her diet should be liberal, her living rooms spacious and airy, and her association cheerful. Wherever possible, she should be free from the anxieties of earning a livelihood or the pompous frivolities of wanton society. The boundless extravagancies of extreme wealth and the awful misery of extreme poverty, both alike, sap the vital forces of the mothers as well as of their offspring. The State, if need be, should provide for the poor expectant mother at least a few weeks' respite from hard work, previous to delivery and also thereafter. We must allay the anxiety of the primipara by assuring her that pregnancy and parturition are physiologic, normal processes, under proper management devoid of perilous complications and sequelæ; and the multipara should be impressed with the fact that miscarriages and at-

tempted abortions are dangerous experiments, one tending to interfere with normal development of the offspring that are to come later (by leaving the uterus in a more or less permanently diseased state), the other actually injuring the embryo or fœtus during the process of growth and development.

Next to antenatal hygiene the judicious management of labor serves as the most important means in the prevention of amentia in the offspring. Judging by the appalling number\* of cases of paralytic amentia following traumatism during birth, and considering the fact that the cases recorded form but an infinitesimal portion† of the innumerable cases that never see light after delivery or survive the injuries sustained but a few days or weeks, there must be something very seriously wrong with the way midwifery is being practised even in the civilized parts of the world.

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\* Lapage reports 25 out of 96 cases of amentia under his observation; Still 26 out of 135; and my own records show 17 cases out of 91.

† In Philadelphia, for example, out of 39,975 births during the year 1911, 2131 were still-born; and according to the annual report of Miss Julia C. Lathrop, out of 300,000 infants under one year who succumbed during 1911, about 30 per cent. did not live to complete the first month of life as a result of prenatal conditions or of injury and accident during birth!

Due allowance, of course, must be made to the fact that in this country the women who furnish the greatest number of births are of foreign birth and bringing up, and owing to mistaken prudery still cling to the custom of their mothers and depend upon ignorant midwives for the performance of the vital function of the obstetrician. But it is high time that each State and Federal Government should put a limit to this much abused "personal liberty" clause and insist upon licensed midwives (after practical examinations) only being permitted to practise obstetrics in order to safeguard the future welfare of the children as well as the mothers—not to speak of the economic benefits to their respective communities. Medical men also ought to awaken to the gravity of the situation, and on the one hand, refrain from the hasty application of instruments in ordinary cases, and, on the other hand, in difficult labors not to hesitate to invoke the assistance of competent obstetricians, who through skilful manipulation might possibly be able to prevent cranial injury, asphyxia, etc., in the infant, which so frequently lead to cerebral diplegia with amenia. Pituitary extract, the most recent addition to the obstetrician's armamentarium, which in



appropriate causes seems to exert an almost magical effect upon inertia uteri, will undoubtedly greatly help to dispense with instrumental delivery and thus diminish the number of amputations due to this cause.

As regard the postnatal care of the infant in the prevention of mental deficiency, let me urge upon physicians ever to remember that their sphere of usefulness does not end with the perfunctory manipulation of the stethoscope or thermometer, nor even with the punctilious elaboration of food-formulas, diet lists or recipes. The mentality, das *Sinnesleben* (the mind-activity, or the senses) of the child should enlist as much of our deliberative attention as its physical condition, more especially when there is a tainted family history or an environment that is conducive toward a morbid mentality. And while it surely is the physician's paramount duty to safeguard the physical welfare of the child, it is no less important for us to guide its mental destiny. Indeed, strength of mind quite often compensates for weakness of the body, while strength of body only very rarely if ever compensates for weakness of mind.

In the mental training of infants we usually



meet with two extremes. One class of mothers keep their infants in a state of "noli me tangere," hidden in the remotest corner of the boudoir, lest it be bewitched by an "evil eye," or, as they say, unhinged by the premature sensitization of the brain; the other class of mothers injudiciously strain their infants' cerebral functions to the breaking point, make them the central figures of attraction of their household, teach them to sing and to recite, to read and to write at a very tender age, when they are barely out of their bottling period. These two extremes in mental training should resolutely be discountenanced, and the happy medium chosen. The infantile brain, like the potter's clay, needs molding while it is fresh and pliable, but it must be mastered skilfully and gently to avoid exhaustion of the brain cells or their disarrangement. The mental training should begin when the infant is about three months old. It should be picked up a few times a day, put on the lap and supported with the forearm, and shown a few lustrous things to stimulate its power of vision and attention. Gradually some objects should be put in its hands to train them to grasp. As it gets a little older, it should now and then, for a few minutes at a

time, and properly supported, be sat up in its carriage or bed, and allowed, as it were, to make a general survey of the beautiful world and the grand things that help to make it so. At six months of age, if strong enough, it should be put in a baby chair, given a few harmless little toys to play with, and be permitted to be accosted by some intimate friends of the family, in order to get the child accustomed to distinguish strange faces. Another few months later it should be gradually taught to perform some simple "baby tricks," such as clapping hands, and the like. In suggesting these procedures I do not at all intend to convey the idea that every kin and friend of the family should be invited to exhaust their ingenuity to devise means to entertain the baby. Quite the contrary, we must ever bear in mind that an infant is easily fatigued, irritated and indisposed, and hence should not be overtaxed even by the simplest methods of training. This holds true also of older children, for many a supposedly nervous, naughty, intractable, listless or morose child, on careful inquiry, is found to be suffering from the effects of overtaxation of its mind by injudicious entertaining, training or education.

On a previous occasion attention has been directed to the serious consequences, in the way of mental affections, not rarely following febrile diseases of childhood. Here also it is in the physician's province to educate the public that measles is not a "children's ailment that every child must go through," the whooping-cough is not "harmless and makes the children fat thereafter," and that "scarlet rashes" are "of no consequence, and the result of a spoiled stomach or teething." And the sooner the people will appreciate that grave danger lurks even in the most benignly appearing attacks of infectious diseases, the sooner will the mental deficiencies arising from this cause dwindle down to insignificance.

Do what you will, even with the most scrupulous application of all the aforementioned preventive measures, amentia, either of prenatal, natal or postnatal origin, will exist as long as man will inhabit this world. We are therefore obliged to suggest some therapeutic measures which will at least ameliorate the condition of these unfortunate human beings. But before going into the details of treatment, it is essential for us to determine how many of them live long

enough and are sufficiently gifted to avail themselves of the benefits of medical and surgical treatment and of the systematic training we are about to recommend.

Genuine congenital idiots are usually short lived. This is true especially of the congenital hydrocephalic, paralytic (associated with porencephaly and cerebral sclerosis), Mongolian with congenital heart disease, and the amaurotic. The great majority of them succumb during early infancy or childhood either to general debility or to intercurrent diseases, more particularly pneumonia and tuberculosis. The pneumonia is usually of the hypostatic variety which readily supervenes after trifling ailments which in any way tend to depress the vitality of these weaklings. The prevalence of tuberculosis among them can often be traced to an hereditary disposition, superinduced by the ever present nasopharyngitis and adenoids, or to direct infection of the alimentary or respiratory tract as a result of the extremely unhygienic habits of the majority of low grade aments. As is well known idiots delight in rolling in filth and chewing on anything and everything picked from dirty floors and streets, and it is quite reasonable to suppose

that these things harbor a multitude of pathogenic micro-organisms, the tubercle bacillus among them. On rare occasions we meet with aments of very robust constitution, who are not susceptible to the ordinary children's diseases, and often, almost for spite, surmount violent attacks of exanthemata, gastroenteritis and the like, even if left alone without proper hygienic care, suitable feeding or medication. This is often true of the microcephalic idiot with the miniature brain and head (see p. 29), and the postnatal cretin.

The mortality in feeble-mindedness of postnatal origin, more especially in children who receive good care and treatment, as a rule, is not much higher than in normal children, provided they remain free from convulsive seizures and are able to be around and about. As regards the future mental progress of aments, each case must be judged individually. As a rule, however, mentally deficient children who at about three years of age are able to make more or less free use of their extremities, in the course of time are amenable to proper training. Some of them as they grow older can be made useful by teaching them light outdoor occupations, such as gar-

dening or to assist in farming; others by learning to help along in different trades, e.g., basket making, carpet laying, carpentry, and others again by doing errands, etc. Under these circumstances, more particularly since errors in the diagnosis of the exact type of amentia dealt with are not at all uncommon even with the most experienced observers, it is hardly just or expedient to declare a case of feeble-mindedness unimprovable without giving it a fair test by way of physical and mental training, or, possibly medical or surgical treatment—whenever there is reason to believe that these therapeutic measures might prove of some benefit to the child, or at least will do no harm.

## CHAPTER V

Active Treatment of Amentia. General Hygiene. Training by "Incentive" Method and Physical Therapeutic Measures. Organotherapy. Thyroid Transplantation. Drugs. Surgical Procedures.

WE can now pass on to the treatment of amentia, and since our essay is intended to be limited to the discussion of mental deficiencies in infants and children under school age, we propose to speak only of therapeutic measures (hygienic, pedagogic, physical, medicinal and surgical), as they are applicable chiefly in the management of aments under five years of age.

### HYGIENE

An ample supply of fresh air and good food, bodily cleanliness and proper clothing are essential prerequisites—in many respects more so in the care of aments than in normal children. Owing to the tendency of mentally deficient children to contract tuberculosis and the frequency of respiratory embarrassment as a result of nasal disease or deformity, they should be kept out-

doors most of the day and in thoroughly ventilated rooms during the night or inclemency of the weather. This fact should strongly be impressed upon those who take care of these children, since most mothers are apt to mistake the cause for effect and attribute the difficult breathing and nasopharyngeal catarrh to "catching cold" in the street.

Several precautions have to be taken in feeding mentally backward infants. Some of them, owing to their voracious appetite and lack of prehension of the sense of heat or proportion, are apt either to burn their mouths with hot food or swallow big morsels and thus permanently impair their powers of digestion. In the majority of cases, therefore, it is required to properly prepare and subdivide the food, just ready for consumption. Others again, because of imperfect sense of taste and inability to manipulate the tongue, often refuse food, especially solids, and have to be fed with small quantities of food at frequent intervals. Owing to nasal obstruction, either as the result of adenoids, nasal deformities or general debility, some congenital idiots experience considerable difficulty in nursing on the breast, and hence it is often necessary to pump



off the breast milk and to feed the baby either through a bottle and small nipple or by means of a spoon. As soon as possible, let us say from the eighth month on, mentally backward infants should be put on a mixed diet, in order to prevent rachitis or scurvy. We usually begin with small quantities of fresh fruit juice, beef juice, strained vegetable soups, coddled egg, cereals with milk, stewed fruit and vegetable purée, gradually increasing the quantity of food as they get older, so that at the age of three or four years they can be put on the following dietary:

On rising in the morning, 4 ounces of milk, preferably boiled.

One hour later, about 8 a. m., half an ounce of orange-, pineapple- or grape-juice; four to six ounces of well-cooked cereal in milk and a little butter, e.g., oatmeal, farina, sago, rice, tapioca, cream of wheat or arrowroot; milk toast and coddled or poached egg.

At about 12 mid-day, four ounces of broth with some cereal or toasted bread; two ounces of vegetables (potatoes, carrots, spinach, cauliflower, beans, peas, etc.) well cooked and finely mashed; half an ounce of finely chopped scraped beef, chicken or lamb chop, or boiled or broiled

white fish. If the child is still hungry we may add a slice or two of stale bread and butter, divided in small pieces.

At 4 p. m. one cup of boiled milk, with a few biscuits, with jelly or butter, or a ripe banana.

At 6 p. m. one coddled egg, bread and butter or jelly and four ounces of boiled milk, or cereal pudding or custard, bread and jam or treacle; or a cup of cocoa with toasted bread thrown in and a small portion of stewed fruit.

Water should be given between meals.

Before and after each meal the child's hands and face should be thoroughly washed as a routine procedure, which may aid also in teaching the child cleanly habits.

The training of cleanliness is very essential to the child's future welfare, since it not only serves to make it more presentable to those coming in contact with it, but, which is by far more important, it acts as the most efficient preventative of diverse local and constitutional infections. In addition to frequent local cleansing of the body as necessity arises, it should receive daily a tub bath, preferably in the evening, in the same manner as normal children. Regardless of the mental condition of the child every effort should

be made to train it to respond to nature's calls. From six months of age on it should be put on a nursery chair at regular intervals, at first every two hours and later every three or four hours. If its bowels do not move spontaneously, after it has been sitting on the chair for several minutes, the infant should be trained to press by inserting into the rectum a small soap stick or glycerine suppository. After persistent training even the idiot will gradually learn to understand what is expected of him, when placed upon the nursery chair, and in time he will of his own accord announce his desire to urinate or defecate. Amients, more so than normal children, should receive more care as regards changing their diapers and keeping the mouth, nose and eyes clean, in view of the fact that, as a rule, they are less sensitive to pain and annoyance, and hence are less apt to complain when those portions of the body are in an irritated state. Moreover, especial attention should be paid to apparently the simplest kinds of cutaneous eruptions, as these often serve as portals of entry to systemic infections.

Mentally deficient infants, especially if delicate and thin, should be very warmly dressed. They may suffer greatly from the effects of cold

and yet fail to appreciate it—owing to dullness of sensibility. Chilblains and frost bites are quite common among them and general circulatory disturbances are frequently encountered, especially in Mongolians and cretins. Flannel and silk underwear should be given preference to cotton or flannelette. During the cold season special protection should be accorded to the hands, feet and ears, and very delicate infants may preferably be kept indoors, in well ventilated rooms, particularly if they show a marked tendency to congestions of the nasopharyngeal mucous membrane.

#### TRAINING AND PHYSICAL THERAPEUTIC MEASURES

The main object of systematic training in amentia is to render the mentally backward child capable of helping himself in the care of his body, to look out for his health and comfort, and later to learn some simple occupation to earn a livelihood. This requires first of all the ability to exercise the voluntary musculature. As in a great many aments several groups of muscles are either atrophied and incapacitated from disuse or actually paralyzed, we must endeavor to estab-

lish or re-establish their functions by passive motion, massage, hydrotherapy, electricity and active exercise. These procedures must be continued uninterruptedly daily for months and sometimes for several years notwithstanding their seeming futility. In the end we are usually amply rewarded by success for our patience and perseverance. Even in the paralytic it has repeatedly been found that the function of the diseased cerebral area has been taken up by the corresponding healthy structures of the brain, and as has been shown by Vitzow, Pfitzner, Merk, Cattani, Klebs and others, in exceptional cases actual regeneration of nerve tissue occurs where its injury is not very pronounced. The physical treatment of the affected limbs should begin as soon as their weakness or paralysis has been determined, regardless of whether or not reaction of degeneration has supposedly taken place. For it is by far safer to err in the direction of overtreatment than undertreatment. Let me illustrate this point by a case under my observation. It concerns a three-year-old boy who received severe cranial injuries during instrumental delivery. The abrasions from the blades of the forceps were still visible six weeks after

birth, when the child came under my care. There was at the time distinct paralysis of the face, of both upper extremities and on the right leg. A few days before, the baby was seen by a noted neurologist who thought there was no hope of it ever recovering, and discouraged further treatment. The case did look hopeless, yet the parents failed to reason as disinterestedly as the learned doctor did, and I also agreed with them that the baby ought to be given a chance to fight for its life. The facial paralysis proved peripheral in character and the paralysis of the left arm was of the Duchenne-Erb type, and both disappeared under massage, electricity and patience, while the right hemiplegia has improved so much that the boy is able to walk about with ease and to participate in all sorts of children's games. Moreover, his mentality seems perfectly normal. I may add, by the way, that his cranial circumference measures about 22 inches, and by its shape and consistency gives the impression of a macrocephalus accompanying hypertrophy of the brain.

There are a few practical points to keep in mind in the application of massage, electricity and hydrotherapy. The massage movements

should consist of stroking, friction, kneading, light pinching, tapping and rhythmic vibration. The duration of each treatment should vary from a few minutes in the beginning up to a quarter of an hour after the child has become used to the manipulations. This should be followed by passive motion of a few minutes' duration. The massage should be gentle, preferably by means of talcum powder, since it allows the hands to glide smoothly over the body surface.

Electricity should be administered from ten to twenty minutes at a sitting, either daily or every other day, using the mildest current that will cause muscular contraction without undue pain. The galvanic and faradic currents, alternated with the sinusoidal, answer the purpose well. If single muscles or muscle groups are affected the sponge electrodes are to be applied near or at the points of origin and insertions of the muscles, while if whole extremities are involved, we apply a large flat sponge electrode, well moistened in warm salt water, on the spine and stroke the affected muscles with a small electrode.

Hydrotherapy is particularly useful in amenia associated with muscular rigidity and general cerebral irritability. A warm (98 deg. F. to 101



deg. F.) tub-bath, of from five to ten minutes, should be given once or twice a day, and while the child is in the tub its limbs should be gently rubbed with a rough flannel and if possible extended and moved in all directions.

Simultaneously with the application of these therapeutic procedures, we employ systematic training of the voluntary musculature and the special senses which are intended to foster the physical and mental development of the backward child. In pursuing this course of treatment we must as closely as possible follow the successive steps taken by nature in the unfolding of human intellectual faculties, and avail ourselves of the child's natural instincts to assist us in our efforts. In our study of the normal baby we have noted that immediately after birth it is endowed with the instinct of suckling, of "fishing" for food, and to cry when hungry or thirsty. This instinct is as strongly developed in the idiot as in the normal child. Now, then, since the struggle for food, for self-preservation, forms the ever and everywhere dominating and propelling force of evolution in the animal kingdom of the entire universe, and has formed the most vital incentive



even in the earliest primitive man to devise ways and means for his sustenance and perpetuation, I believe that this irresistible force to quench thirst and to appease hunger ought to be sufficiently powerful to awaken even the total idiot from his mental torpor, and to induce him, as it were, to struggle for his existence. Indeed, the longer I practice this *incentive* method of training of the mentally defective infant, the more convinced I am of its superiority over every other method of training in vogue. We make use of his desire for food to teach him how to look, how to listen, how to pay attention, how to grasp, how to imitate personally and with objects, how to walk and how to talk—all in the order in which the normal baby acquires these faculties, except, of course, at greatly delayed periods as compared with the age of the normal baby. The sooner the training is begun, the more promising are the results, principally because in amentia of long standing the brain cells usually entirely lose their regenerative quality. This fact should strongly be impressed upon the unfortunate parents who rarely note any mental deficiency in their infants, and

if they do, are often led to believe that they would "outgrow their weakness when they get to be seven years old."

As amentia is very readily recognizable in an infant about six months of age we proceed with the training in the following manner:

1. If it is a nursing baby it is put on the lap facing the mother or wet nurse, and after exposing the breast the baby's hands are brought in contact with it and manipulated so as to make them grasp it. This is repeated for a few minutes before each nursing. We next bring the baby near the breast nipple and squirt a little milk in its mouth. As the baby puckers its lips to grasp the breast nipple, we promptly pull the child back, so as to force it to struggle to get a good hold of it. This also is repeated for a few minutes. If it is a bottle-baby, we perform the same manœuvres with the bottle and nipple. As it gradually learns to recognize the bottle, we next endeavor to train the baby to follow the course of the bottle, by slowly moving it before its eyes in all directions, before allowing it to get a hold of the nipple as it comes near its mouth. It may take several days, weeks or even months to accomplish this trick, but patience and persever-

ance are the key-notes to success in the training of the mental defective.

2. After the baby has acquired the power of grasping the bottle, we place the child in semi-recumbent posture with the head resting on a small pillow, put the bottle in its hands, and as it is about to start to suck the nipple, we slowly pull the bottle backward and continue to do so while the child is making every effort to bring the bottle to its mouth, and by hanging on to the bottle lifts itself from the recumbent to a sitting posture. This up and down movement is exceedingly useful to strengthen the arm and spinal muscles and to train the child to raise himself from a recumbent posture. Of course, these exercises must be continued for several minutes before each feeding.

3. Like normal babies aments also should receive a mixed diet when they reach eight or nine months of age. Most of them, as a rule, can be readily induced to take eggs and fruit, which are most excellent bone and brain builders. We hold the egg in front of him and feed him with a spoon at short intervals, making him wait for the next spoonful until he shows anxiety to get it. If a white egg does not attract his attention

we color it red or blue, and move it in different directions to teach him to follow objects. Similarly hold a red apple in front of him, scrape some of it and feed him at short intervals; bring the fruit near his nose and let him learn to perceive its odor. Some idiots have a highly sensitive sense of smell, and by using attractive odors as a bait they can be induced to awaken from their apathy and to better respond to the systematic training. Next place the child near the side of the crib and put its hands on the upper crossbar, and while you hold the apple at a short distance above its head, with one hand, help the child lift itself from its position with the other hand; as he accomplishes it, let him have some of the apple (orange or peppermint stick) and let him go through the same performances again and again to earn some more of it.

4. By means of a rod and cord suspend a red apple or orange in front of the baby, and let it hang there for a little while. If he remains passive bring the apple near him and let him grasp, smell and taste it; if he is now attracted by it swing the apple to and fro and encourage him to follow it with the hands and grasp it. Repeat the exercise several times and let him have some

of the apple for each successful effort. Toys may be used instead of eatables where the child shows preference for the former. By continuing these exercises the child gradually gains considerable power of attention and muscular coordination.

5. Aments should early be taught to feed themselves. Sit him in a baby chair, if need be, well supported with pillows, and place the food before him. Give him a taste of it, and if he is good and hungry he will "fish" for more. In this event, if the food is solid (e.g., a zwiebaek) put it in his hand and guide him repeatedly to bring it to his mouth. It is usually a very difficult task to teach mentally deficient children to feed themselves, but hunger and persistent training will accomplish it in the end.

6. To teach him to stand we place him against the side of the bed with his arms crossing the top bar, and feed him with a spoon in such a manner that he is forced to raise his head to receive the food. In the beginning it is usually required to support his back to keep him from falling. As he learns to stand, put him in a softly padded walker, the top of which snugly surrounds his waist. Keep the dish of food in front of him, give him a mouthful of it and take a short step

backward; let him follow you (which in the beginning may call for your assistance); give him another mouthful and let him again push forward. Repeat this a few times a day, at first only for a very short time, in order not to tire him. A doll carriage may occasionally suffice as a support instead of a walker, especially after the child has partly learned to walk, and only needs additional exercise.

7. Gradually train him to walk without any support. This is best accomplished first by standing him against a wall, and while facing him let him grasp one of your index fingers, and follow you while you take single steps backward. Later extend to him a cane or rod instead of your fingers. If he hesitates to follow you, use some fruit, sugar or candy as an incentive, which you hold in front of him and reward him with for successful efforts. Or put his food on a low table, direct his attention to it and lead him toward it. In time he will go after his food without being led.

8. All the while the child is receiving instruction keep on telling him what you are doing and what you wish him to do, regardless of whether or not he understands you. Gradually he will

learn to understand at least part of what you are telling him. Use single words instead of sentences, e.g., eat, drink, walk, etc., and repeat the words in a firm tone of voice, in order to make a lasting impression upon the auditory center.

9. After he has learned the different exercises, you can begin to interest him in drills, tricks, and games as practiced in modern kindergartens. Almost all mentally deficient children are charmed by music, it is therefore of great advantage to make use of harmonious strains of the piano to arouse the dull from his slumber and to sooth the discordant impulses of the agitated child. Different strains of music should be used for different sets of actions, in order to train the child's auditory apparatus to connect the particular melody with the particular act he is to perform—in other words, one and the same melody with his meals, another one when he marches, a third when he plays a certain game, etc. Music should be employed also in training him to speak. Thus, while playing the piano sit the child in front of you, attract his attention to your mouth, and with a tone of voice corresponding to the strains of music keep on repeating single syllables or words, e.g., ba-ba, for one melody; la-la, for



another; ta-ta, for a third and so forth, gradually lengthening the syllables into whole words. In speech, as in other exercises, its accomplishment is often facilitated by using food as an incentive, i.e., give him a piece of candy every time he makes an earnest effort to pronounce certain syllables or words.

10. Imitation is the mother of experience. Teach him to imitate your personal movements, such as kneeling, sitting, standing, opening and closing of the mouth or hand, throwing a ball or catching it, and similar exercises.

11. Sit the child near a table facing you; spread out in front of him some candy or other eatable he is fond of, and let him taste some of it. If he likes it, he will surely look for some more of it. Now cover the remaining pieces of candy with a strip of paper, leaving part of it exposed. If he shows ability to remove the paper and to help himself to the candy, put the latter in a little box, first without a cover and then with a transparent cover. Now teach him to uncover the box, and if he succeeds doing it reward him with a piece of candy. Next put the candy in a more complicated contrivance, and the more ability he shows to help himself, the more diffi-



cult should it be made for him to find the thing he is looking for. By repeated training he will gradually learn to help himself in many other respects.

12. Place the child in front of a step ladder and put the candy on one of the rungs and encourage him to reach for it. Of course, at first he will need your help. As he ascends the ladder, hand him a piece of candy. Repeat this manœuver again and again, and as he succeeds in getting there, elevate the box to a higher level. In a similar manner reverse the performance, i.e., make him descend in order to reach the box. As he learns to accomplish this with ease, teach him to climb stairs, first by supporting himself with the hands and later by doing it without support.

The performances, of course, can be multiplied almost *ad infinitum*. But there are two essential ideas ever to be kept in view in the training of aments, viz., First, no coercion or force is to be applied; second, no time and energy should be wasted on exercises which are not absolutely indispensable to his welfare. If we succeed to train a deficient child under five years of age, to be clean, to feed himself, to

walk, to understand words spoken to him and possibly to make himself understood, even if only by single words, enough indeed will have been accomplished. By opening up the avenues of approach to the dormant, deficient infantile brain, the brain itself will spontaneously evolve its resourcefulness, round out the experience, and receive new impressions.

In suggesting the aforementioned exercises I presume, of course, that we are not dealing with total idiocy accompanying extreme degrees of hydrocephalus, microcephalus, diplegia, etc., or amaurotic family idiocy. In these cases no amount of conscientious training ever will bear fruit in restoring degenerated brain tissues to normal function. Moreover, the span of life of these unfortunates measures but a few years. On the other hand, when confronted by a mentally deficient child that is free from gross cerebral lesions and shows some response to outside influences; for example, a child of two years responds to the mental tests of a normal baby of six months, painstaking and persistent training will most assuredly bring forth very gratifying results, even though in the beginning nothing but failure will seem to crown our efforts.

The late Edouard Séguin, one of the early pioneers in the training of mental defectives, when once asked why he kept on repeating the same movements a hundred times a day, he replied because the child does not make them right ninety-nine times. This was the secret of his phenomenal success. And it is essential to impress upon the parents that unless they themselves are endowed with an ample supply of patience, tact and perseverance, to keep on teaching their child the same things for days, weeks and months, this work should be intrusted to some one who possesses these qualities, or no great achievements need be expected.

Finally, in training the weak-minded it is well to remember that the unfortunate baby is not to be blamed for his failure promptly toprehend and to copy the apparently simplest rudiments of thought and action. He is heartily to be pitied rather than disdained, and as the great majority of aments are providentially blessed with a content and joyful disposition, we might as well refrain from shattering their peace of mind by undue harshness or rough handling.

## ORGANOTHERAPY

When discussing cretinism attention has been directed to the marvelous physical and mental transformation occurring on the administration of thyroid extract in amentia due to thyroid insufficiency. Wherever the cause of amentia is uncertain, I made it a rule to give the child the benefit of a few weeks' thyroid treatment in order to determine whether or not the thyroid was at fault. Moreover, for the last two or three years I have frequently found it of advantage to supplement the thyroid medication by the extracts of parathyroid, thymus and pituitary glands, in accord with the established fact that whenever the thyroid is affected, the functions of the other glands are also more or less impaired. In one case particularly the effect of the combined glandular medication was singularly striking. It concerned a six-year-old boy who for three years had been treated with thyroid extract by several eminent clinicians. When I first saw him he measured 35 inches in height and weighed 41 pounds. His voice was husky, and he could pronounce but a few words in a draggy, staccato sort of fashion. His face and lips were edema-

tous, and his tongue protruded slightly. When led by the hand he was able leisurely and awkwardly to move along, but if left alone he was barely able to take a few steps without stumbling. I put him on the aforementioned glandular-extract-compound, and he gained  $1\frac{1}{2}$  inches in height, three pounds in weight and a great deal in intelligence. He became so active that the mother experienced considerable difficulty to "restrain him from following the boys in the gutters." As until he came under my observation he had regularly been receiving from two to five grains of thyroid extract daily without appreciable benefit, I could not help but believe that the marked improvement in his condition was due solely to the addition of the parathyroid, thymus and pituitary extracts. The mode of administration of the glandular extracts varies somewhat with the age of the child and the degree and duration of the affection. In congenital cretinism I order one grain of thyroid powder twice a day to a bottle-fed baby and only half the quantity to a breast-fed infant, having observed that the latter develop the manifestations of cretinism more slowly than the former—owing probably to the fact that the breast baby in the

first few months of life receives some thyroid through the mother's milk. If the case fails to show improvement in about four weeks, I add to each dose of thyroid a quarter of a grain each of parathyroid, thymus and pituitary extract. This represents the usual dosage for an infant up to one year of age. Older children should receive half a grain of the thyroid powder and a quarter of a grain of each of the other glandular substances for every additional year of their respective ages up to five years. The dosage in tablet form is about twice as large as that of the powder. After considerable improvement has taken place in the child's general development, the dose of the thyroid compound should gradually be reduced to once a day, once every alternate day, and every third day. Where organotherapy gives rise to cardiac palpitation, undue restlessness, or gastric irritability, the treatment should temporarily be suspended until the toxic symptoms have disappeared, when the medication should be resumed in smaller quantities, and gradually increased.

A very valuable suggestion as regards thyroid therapy has been offered by H. Stern. Finding that human thyroid contains about sixteen times

as much arsenic as the sheep's thyroid, he recommends the addition of minute doses of arsenic, preferably in the form of sodium cacodylate, to the thyroid gland substance. Thus combined, the activity of the thyroid is greatly reenforced, allowing a considerable reduction in its dosage.

#### THYROID TRANSPLANTATION

More recently a renewed attempt has been made to cure cretinism and myxœdema by means of Thyroid Transplantation and the following report (*Jour. A.M.A.*, June 6, 1914), which is a brief summary of the papers on the subject read at the annual meeting of the German Surgical Society, gives a general idea of the success thus far obtained.

Kocher, of Bern, reported the permanent results of thyroid transplantation in man. In animal experiments it was possible to transplant the thyroid gland and its vessels, but only in case of auto-transplantation, which is of no significance for the present question. No success has hitherto been obtained by means of homotransplantation. Probably the question also will be solved by some method of weakening the immunity of the recipient and increasing the vigor of

the transplant. For the prevention of tetany, when it was not possible at the operation to preserve the epithelial bodies, Kocher immediately reimplanted these. Also for the prevention of cachexia strumipriva, when for any reason the entire thyroid had to be removed, he reimplanted a portion of the healthy tissue. In cases of cretinism, in which the patient's own thyroid is not available, the attempt must be made, if necessary, to obtain material from relatives. The successful cases which Kocher has to show have not been controlled by histologic examination because he does not regard himself as justified in removing again a gland that has once been implanted.

He plants the graft in the bone-marrow, the peritoneal cavity, the preperitoneal tissue, the omentum or, following the method of Payr, in the spleen, which seems to him especially suitable for the purpose. In ninety-three cases he had eighteen favorable results; some of the patients had previously been taking thyroid preparations internally. It should be noted further that there are cases in which these preparations are tolerated only after a thyroid-grafting operation. In cases of cretinism, he had good results in twenty-



one cases. For transplantation, he prefers the thyroid from a patient with exophthalmic goiter which is in the stage of hyperthyroidism. For good results, a single transplantation is not sufficient; new material must constantly be supplied either by the administration of thyroid preparations or by multiple transplantations. In cretins, moreover, preliminary long-continued administration of the thyroid is necessary for success with the transplantation.

Eiselsberg, of Vienna, reported a case of myxœdema in which an improvement was secured by repeated transplantation; but after a certain interval a new transplantation always became necessary. He has succeeded in transplanting the epithelial bodies but with only a temporary effect.

In a case of congenital lack of the thyroid with complete idiocy, which Payr, of Leipzig, reported, an astonishing and unprecedented improvement lasting for two years and four months was achieved by transplanting a large piece of the mother's thyroid; but later a complete retrogression occurred. Three other transplantations gave satisfactory results. Payr said that in cases of acquired imperfect function of the thy-

roid, after failure of thyroid treatment, transplantation may be undertaken with good prospects of success. The causes for failure of transplantation of congenital cretins are partly the severe disturbances of development in the infantile brain which are present at the same time and partly the necessity of taking the graft from another individual. For an early diagnosis, Payr recommends a small exploratory incision in the neck. In this place, then, the transplant itself may be inserted or it may be embedded in the capsule of the thymus. There are many differences between the mode of action of transplanted thyroid tissue and thyroid treatment with the dried gland.

Stieda reported three cases of pronounced myxœdema with more or less evident idiocy, in an eight-year-old boy, his sister of seven and an eight-year old girl. After the implantation of thyroid tissue pronounced improvement was evident in all three cases. At first it showed rapid progress and then it became stationary. A disappearance of the myxœdematous changes in the skin could be demonstrated as well as growth in height and an evident increase in intelligence. Two of the children are still under observation,

but the third died nearly four years later, after an operation for tuberculosis. At the necropsy this case, which had been especially improved showed that microscopically the thyroid tissue implanted in the head of the tibia was no longer recognizable but, on the other hand, a small thyroid gland was found which had escaped discovery during life. Aside from the grafting operation, the children received no other treatment.

Schaack, of St. Petersburg, has repeatedly undertaken the implantation of the thyroid in the bone-marrow or in the subcutaneous adipose tissue and has seen improvement which as a rule, however, lasted only two months. He has never observed a permanent cure.

Müller, of Rostock, implanted the thyroid in the bone-marrow of the tibia in one cretin, with merely temporary improvement. He later opened up the tibia again but found nothing left of the thyroid gland except a few isolated cells.

Enderlen, of Würzburg, stated his conviction based on wide experience that the thyroid implanted in this way will not long maintain its life.

## MEDICINAL TREATMENT

There are several other medicinal preparations which have to be resorted to in the management of the different forms of amentia. Regardless of cause, it is often judicious to place an ament on a thorough anti-syphilitic treatment, more especially, of course, when the Wassermann reaction is positive or there are reasons to suspect syphilis either from the history of the case or appearance of the patient. Opinions are still at variance as regards the advisability of employing neosalvarsan in the treatment of syphilitic amentia in children, and I am inclined to give preference to mixed iodid and mercury treatment, unless there be special need for hasty action, e.g., syphilitic hydrocephalus with marked intracranial pressure. If neosalvarsan is indicated it should be administered either intravenously, intramuscularly or per rectum. The usual dose is from 2 to 5 grains dissolved in freshly prepared distilled or sterile water, made ready for immediate use. The effect of the neosalvarsan should be controlled by the Wassermann reaction, a second dose being administered after two weeks if necessity arises.

The iodids are indicated even in the absence of a syphilitic taint, acting as they do as powerful alteratives and eliminants of diverse systemic poisons. One grain of the sodium iodid, twice daily, for every year of the child's age, is ample for ordinary purposes. The sirup iodid of iron, in ten-to-twenty-drop-doses, may be alternated with the sodium iodid, or some of the newer iodid preparations may be used instead. To obtain results the iodid should be continued for several months, with occasional intermissions of short duration, in order to avoid gastric irritation. It is often found very beneficial to combine the sirup iodid of iron with cod-liver oil and the sirup of lime hypophosphites, more especially where rachitis complicates the amentia. General tonics and appetizers are almost always indicated, for we hardly could make proper use of our *incentive* method of training, when the incentive, hunger, is lacking. Small doses of the tincture of nux vomica and cinchona compound, in orange sirup, before meals and dilute hydrochloric acid and essence of pepsin after meals act exceedingly well both as tonics and digestants and should be prescribed as necessity arises. In paralysis or general muscular debility it is

often advisable to administer strychnine by mouth or even hypodermatically. One-three-hundredth of a grain for every year of the child's age up to five years will ordinarily suffice in cases of moderate severity. The dose may be repeated twice or three times a day. Sometimes sedatives are indicated, and I have found that small, frequently repeated doses of codein, dionin or heroin, act very much more promptly in relieving attacks of twitching, extreme restlessness and insomnia, than the bromids or other hypnotics. Of course, in epilepsy, the bromids are indispensable, and instead of, as is usually advised, giving small gradually increased quantities of bromids, I have found it very much more profitable to start with large doses, and to reduce them, after the periodic attacks have been arrested. Thus, for every year of the child's age I give one grain each of potassium, sodium and strontium bromid, three times a day, and continue the same until I have succeeded to arrest the usual fit for several months. Then the dosage may slowly be reduced if found that the child is too drowsy or signs of bromism make their appearance. I prefer to combine the bromids with small doses of Fowler's solution and

the mixture of rhubarb and soda—the arsenic seemingly preventing bromism while at the same time acting as nerve tonic, and the rhubarb and soda serving to subdue the undue gastric irritation. In habitual constipation which is the rule in mentally deficient babies, phenolphthalein, gr.  $\frac{1}{2}$ , or aromatic fluid extract of cascara sagrada, ℥. 10 for every year of the child's age will be found efficient, particularly if the movement be started with a small injection of warm soap water or glycerine suppository. In very young infants milk of magnesia (one or two teaspoonfuls, best mixed with the entire 24 hours' quantity of milk) usually answers the purpose. Especial attention should be paid to the nasopharynx. Where adenoids exist and greatly interfere with respiration, they should be promptly removed, otherwise they can be kept from doing much harm by keeping the nasopharynx clean with Dobell's solution and adrenalin (1:1000) equal parts and the instillation in each nostril of a few drops of a 25 per cent. solution of argyrol, twice a week, until the inflammation and hypertrophy of the adenoid tissue has considerably subsided. Intercurrent diseases, of course, must be treated according to indications in the same manner in



aments as in normal children, except that greater attention must be paid to the prevention of passive, hypostatic pneumonia, even with trifling ailments which ordinarily are entirely free from complications in normally developed children.

#### SURGICAL TREATMENT

Surgery as an aid in the cure of idiocy and the allied mental deficiencies has been resorted to especially during the last two decades. The results obtained, however, are far from being satisfactory, except in cases of paralytic amentia due to cerebral compression, where early decompression has not rarely brought about complete regeneration of the brain tissues involved and *restitutio ad integrum*. Harvey Cushing has performed quite a number of craniectomies on the newly born to relieve cerebral compression resulting from intracranial hemorrhage during birth (which, as previously mentioned, forms the cause of amentia in about 30 per cent. of the cases on record), and is of the opinion that with proper regard of hemostasis and careful avoidance of undue exposure, the newly born will stand a cranial operation well, its life will often be saved, and in many instances develop normally.



Roswell Park maintains that where a reasonable integrity of brain structure can be assumed, there is no reason why craniectomy or decompression should not be given an opportunity in imbecility and psychic disturbances, in order to relieve pressure and permit more normal development. And William Sharpe and H. F. Farrell claim very good results from cranial decompression in cases of spastic paralysis (with mental deficiency) of hemiplegic, paraplegic, and diplegic type with a *definite history of difficult labor with or without the use of instruments, in which on ophthalmoscopic examination signs of intracranial pressure are shown in the dilated retinal veins, and a blurring and haziness of the optic disks, especially of their nasal halves.* In these cases they perform a large right subtemporal decompression, and if the intracranial pressure remains high, they perform a left subtemporal decompression the following month. The after-treatment, which is of very great importance, consists in the correction of deformities by tendon lengthening and stretching of the contracted muscles; the maintenance of the corrected positions by the employment of especially adapted and properly fitting braces, and skilled

massage in conjunction with short applications of galvanism and faradism. A careful, systematic course of muscle training is carried on daily. They claim marked improvement not only in the spasticity but in the mental condition of the patients as well, so much so that they are able "to receive the co-operation of the patient in the carrying out of the after-treatment." Of course, the earlier the decompression is performed, the greater the opportunity and facility for the compressed brain structures to adjust themselves in their normal relations and to regain their normal functions.

As has already been stated, the results from operative interference in the other forms of amentia are, to say the least, but temporary. Lannelogue's craniectomy for the relief of the microcephalus, which at first was hailed as a great success, soon proved a total failure and has rightly been abandoned even by its most enthusiastic exponents. Several operations have recently been proposed for the cure of hydrocephalus, but it is too early to arrive at correct conclusions regarding the improvement in the child's mentality and its permanency. Of these operations I may mention G. Anton's efforts to

relieve intracranial pressure by puncture of the corpus callosum\* and Irving S. Haynes' method of treating hydrocephalus by cisterna-sinus drainage. The steps of this ingenious operation are as follows: An incision is made one-fourth

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\* Stieda reported the results which had been obtained at Halle clinic with puncture of the corpus callosum in genuine epilepsy and idiocy. One case of Jacksonian epilepsy with a congested papilla was of special interest in that the attacks gradually ceased completely with no further signs of choked disk to date, after four and one-half years, while in most cases of epilepsy there is only a temporary cessation but a decided weakening of the attacks.

An earlier operation will surely give better results, as the puncture of the corpus callosum is a minor operation, quickly made and not dangerous. It can be done without injuring the brain and without shock, and under local anesthesia. In comparison with the other methods of operating for genuine epilepsy, puncture of the corpus callosum is not only a competitor but probably deserves preference.

Hildebrand of Berlin has performed puncture of the corpus callosum twice and was not very much pleased with the results. He does not regard the operation as so very insignificant, as it is easy to injure a vein and produce dangerous bleeding.

Tilman of Cologne in cases of epilepsy first determines by a lumbar puncture whether there is increased pressure. If positive, he makes a puncture of the ventricle; if he also finds here an increased pressure he punctures the corpus callosum.

Lossen of Cologne applied puncture of the corpus callosum in three cases of hydrocephalus without results. In his opinion, the operation must be made in several places in order to secure success. He recommends puncture of the cisterna magna, but in addition he makes a puncture of the corpus callosum and of the ventricle.

Schloffer of Prague has performed puncture of the corpus callosum about twenty times and observed a temporary unilateral paralysis following it in one case. (*Jour. A. M. A.*, June 6, 1914.)

of an inch at the left of the mid-line from a point about three-fourths of an inch above the margin of the foramen magnum to the same distance above the inion, and the skin with the periosteum reflected to expose the occipital bone. A three-eighths of an inch button of bone is removed by a DeVilbis trephine midway between the margin of the foramen magnum and the inion and from this a gutter, half an inch wide, is cut away to the last point, exposing the dura over the cisterna magna and, in the upper part of the area, the lateral (left) sinus. A suture of vaselin sterilized silk is passed through the dura and arachnoid so as to enclose a space about one-fourth of an inch square, within which space the short arm of the cannula is to be passed. An incision, 1 mm., is now made in the lateral sinus. Into this small opening the long arm of the cannula (previously sterilized in vaselin) is inserted. The fit between the cannula and incision should be very snug, so as to prevent leakage of blood. Another incision, a little less than one inch distant from that just made in the sinus, is then made through the dura and arachnoid into the cisterna magna, in the center of the area previously encircled by the silk suture. Into this

incision is now inserted the short arm of the cannula, and the retention suture is tied across the tube. The cannula may be of silver with an internal diameter of  $1\frac{1}{2}$  mm.: its long arm about 1 inch and the short arm  $\frac{1}{4}$  inch, each end obliquely sharpened; or rubber tubing of firm consistency may be used instead. The shorter the tube the better. Though antedated by Gartner in the basic conception of this operation, and by Payr in an attempt to accomplish it, Haynes' modified operation is much simpler in the technic of execution and less shocking to the patient. Haynes has thus far performed the operation on twelve cases of hydrocephalus. Three of these have "recovered." While these were under observation there was an appreciable improvement in their mentality; it was difficult to tell, however, whether the mental improvement would continue and be permanent in character. The operation should be undertaken as early as possible, preferably before destruction of brain tissue by the excessive intracranial pressure has taken place.

In recommending operative interference for the relief of congenital or acquired physical defects complicating amentia and for the eventual

restoration to normal mentality, considerable conservatism, of course, should be exercised in the proper selection of the cases. But, whenever in our judgment the case in question is entirely hopeless if left alone, and there is the remotest chance, through surgical interference to relieve the idiot of his lifelong misery, we should not at all hesitate to recommend surgical treatment, notwithstanding the accompanying appalling mortality. However, before resorting to surgical intervention, the mental defective should for a reasonable time be given the benefit of some of the other therapeutic measures here suggested in the treatment of idiocy and the allied mental deficiencies in infancy and early childhood.

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## INDEX

- Adenoids, in relation to amentia, 18, 107  
Alcoholism, parental, as a predisposing cause of amentia  
in the offspring, 11; faulty environment, 112  
Amaurotic family idiocy, 77; juvenile form, 79; illustra-  
tive case, 82  
Amentia, general pathology, 2; paralytic; vascular, toxic;  
epileptic, inflammatory and meningitic, 64  
Asphyxia neonatorum, as a cause of amentia, 16, 129  
Ateleiosis, 104  
Athyreosis, 91  
Attention, abnormal power of, 36
- Brain, lesions in amentia, 2; hypertrophy, 58  
Brissaud's type of infantilism, 103
- Cisterna sinus drainage, in the treatment of hydrocephalus, 173  
Clothing, suitable, 141  
Consanguinity, as a cause of amentia, 12  
Corpus callosum, puncture of, in amentia, 173  
Cretinism, 91; differential table of, 88; illustrative cases, 97
- Darwin's theory of hereditary transmission of characteristics, modified, 4  
Deafmutism, in amentia, 13, 109  
Decompression, cranial, in the treatment of amentia, 170  
Diplegia spastica infantilis, with amentia, 67
- Electricity, its mode of application, 145  
Environment, faulty, productive of mental deficiency, 112  
Epilepsy, with amentia, 64; arrest of convulsions, 168; fol-  
lowing natal and postnatal traumatism, 10, 16  
Eugenics, 123  
Exanthemata, in their relation to amentia, 17, 65

- Febrile affections, as cause of amentia, 16, 65; importance of their prevention, 133
- Feeding of aments, 139
- Germplasm, tainted, in its relation to amentia, 9
- Head, measurement, 24; dome or sugar loaf, 18, 51
- Hearing, defective, 34, 109; normal, 28
- Height of normal babies, 25
- Hemiplegia, in connection with amentia, 66
- Hereditary transmission, *modus operandi*, 8
- Heredity of characteristics, permanent, 5; temporary, 6
- Herter's infantilism, 104
- History, personal and family, 19
- Hydrocephalus, 54; illustrative cases, 58; surgical treatment of, 173
- Hydrotherapy, mode of application, 145
- Hygiene, of the child, 137; of the mother, 125
- Hypertrophy of the brain, differentiated from hydrocephalus, 58
- Hypothyreosis, 91
- Idiocy, classification, 2, 46
- Idiotic grunt, 51; status, 36
- Incentive method of training, 146
- Infantilism, 102; illustrative cases, 105
- Intelligence, defective, 41; normal, 28
- Labor, judicious management of, 128
- Literature, 176
- Little's Disease, 67
- Lorrain's type of infantilism, 103
- Macrocephalus, 58
- Malformation, congenital, in connection with amentia, 25
- Malnutrition, its relation to amentia, 18, 111
- Massage, its mode of application, 145
- Measurements of head, chest and extremities, 24
- Medicinal treatment of amentia, 166
- Mental tests, 42

- Mentality, defective, 33; normal, 28; retarded, 107  
Microcephalus, 47; illustrative cases, 51  
Microsomia, 104  
Mongolism, 84; illustrative case, 89  
Moramentia (see Retarded Mentality), 107  
Motion, voluntary, defective, 39  
Music, in training of aments, 153  
Myxidiosis (see Cretinism), 91
- Nanosomia, 104  
Neoplasms, cerebral, resembling anaurotic idiocy, 80
- Organotherapy, 157  
Oxycephaly, 51
- Pain, abnormal sense of, 35  
Paralytic amentia, 64  
Paraplegia, spastica infantilis, 67  
Parathyroid extract, in amentia, 160  
Pathogenesis of amentia, in general, 2  
Perception, abnormal power of, 38  
Pertussis, as a cause of amentia, 16  
Physical therapeutics, 142  
Pituitary extract, in amentia, 160; in obstetric practice, 129  
Pregnancy, judicious management of, 122  
Prohibition, in relation to mental degeneracy, 12
- Rachitis as a cause of mental backwardness, 111  
Retarded mentality (see Moramentia), 107
- Smell, abnormal sense of, 34  
Speech, defective, 40; normal, 29  
Status idioticus, 37  
Stigmata of degeneration, 25  
Surgical treatment of amentia, 170  
Syphilitic, amentia, 69; hydrocephalus, 63; parental, as a cause of amentia, 9
- Taste, abnormal sense of, 34  
Temperature sense, abnormal, 35

- Tests, mental, 42
- Thymus gland, defective in infantilism, 102; in the treatment of amentia, 160
- Thyroid, deficiency, 91; feeding, 157; transplantation, 161
- Touch, abnormal sense of, 35
- Toxemia, maternal, productive of amentia, 15
- Training "incentive," systematic, 146; faulty, 116; of normal baby, 130
- Transplantation of thyroid gland, 161
- Traumatism, antenatal, natal and postnatal, as a cause of amentia, 16, 128
- Tuberculosis, parental, productive of amentia, 22
- Uterine disease, in mother, as a cause of amentia, 13
- Vision, defective, 33, 109, 119; normal, 28
- Weight, of normal babies, 25
- Z-shaped deformity of hands, in Little's Disease, 67

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