*Inaugural Issue*

# PST

# PEDIATRIC SURGERY IN TROPICS

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Platinum Open Access Journal

**Transformative Milestones in Pediatric Surgery**

**Sameh Shehata**
Former President, WOFAPS

**Why We Need a New Journal**

**Vivek Gharpure**
Chief Editor

# Pediatric Surgery in Tropics

January - March 2024 Volume 1: Issue 1

# About the journal

### *Aims and objectives*

Pediatric Surgery in Tropics (PST) aims to improve scientific communication among pediatric surgeons of tropical countries. It is intended to be an author friendly journal. The journal focuses more on the content of manuscripts rather than on their presentation or formatting. We understand the compelling circumstances in which tropical doctors are working. Therefore, we sympathetically accept to publish even partial evidences when advanced facilities for complete work-up are not available. Socio-cultural milieu, economic inequality, limited resources, illiteracy of patients and lack of advanced training that are typical of tropical countries will be taken into account while accepting articles for publication.

### *What do we publish?*

We publish all types of scientific manuscripts that are useful to practicing pediatric surgeons. We publish not only diseases peculiar to tropical countries but also on all aspects of pediatric surgery pertinent to tropical or resource poor settings. We are not limited to article types such as original articles, review articles, case reports, images, letters and viewpoints. Rather we consider every article as a useful scientific communication. Thus we publish even unconventional category of articles. The smallest article we publish is 'clinical tips' of 50-100 words and the largest manuscript is review articles of several thousand words. Opinions regarding legislations and policies that may affect the practice of pediatric surgery will also be considered. We also publish patient perceptions that may provide insightful information to doctors.

### *Copyright*



### *Economic Model*

PST is an e-journal. It is managed on frugality principles. We avoid printing the journal, thereby saving expenditure and remain eco-friendly. Print copies are made available only on special request. All the editors and reviewers offer voluntary free service. The minimal cost of running the journal is met out by charity donations and advertisements. PST is a platinum open access journal that is made available free of cost to all individual readers except institutions and libraries. We neither charge our authors; nor pay them any royalty.

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# Pediatric Surgery in Tropics

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Pediatric Surgery in Tropics (PST) is an author friendly journal. Keeping in mind the limitations of authors from developing countries, we do not strictly look into the manuscript formatting. Rather we only see if there is a clinically useful message in the manuscript. Hence, we request our authors to concentrate more on the content rather than on the format. Those who prefer to use templates (available in PST website) may use it; other may simply type their text in plain Microsoft word or RTF file and send it by email attachment to the editor: ***editor@pediatricsurgeryintropics.com***

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1. Title of the paper
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Each manuscript should include the following details:

- Why was the paper written (introduction, background, problem statement)
- What was done (methods and materials)
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- References to support the authors' statements

We do not bother our authors with stringent word limitations. We only expect them to write briefly and succinctly without losing the readability and usefulness of the text. Number of references should also be kept to minimum necessary number, but sufficient enough to support statements. If the articles with useful message are boringly lengthy, our editorial team will suggest places where trimming is required (Author compliance with editorial suggestions is a precondition for final acceptance).

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During initial submission all figures and tables can be incorporated at appropriate places of the text file and the entire manuscript is submitted as a single word file. After peer review, we may ask to provide separate figure files if required for better clarity of reproduction. In such case, figures can be submitted as JPEG or TIFF files. Resolution must be more than 300 dpi.

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Pediatric Surgery in Tropics
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Editorial

# Why Do We Need A New Journal?

Vivek Gharpure

*Chief Editor, Pediatric Surgery in Tropics*

**P**ediatric surgery and pediatrics as practiced in tropical countries is different from that practiced in developed countries. High population pressure, poverty and constraints of financial support for research limit basic science research in these areas. Most of the clinicians, rather because of large patient load, can only perform clinical research. Countries like India have a large number of independently practicing pediatric surgeons, who have no research support or grants. Pediatric surgery practiced in institutions, teaching or otherwise, is different from pediatric surgical practice in standalone, independent hospitals or nursing homes. Clinical research is poorly represented in traditional medical journals, where emphasis is on basic science research, experimental research or laboratory based research. Technical innovations developed by independent surgeons are inadequately published, though they can significantly alter and improve patient care. Not everything has to be store-bought; many things - instruments, techniques and tools - can be improvised, with the famous principle of *'Jugaad'* (innovation). Therefore, a special journal devoted to high quality clinical research in pediatric surgery from tropical countries will be useful to provide a platform for clinicians in these areas, and expose other clinicians to the kind of work being done there.

This journal aims to provide space to independent surgeons, who work alone, without institutional support, research support and still are able to provide medical and surgical help to a large number of children in need.

The current publishing cycle is overly laborious, prolonged, almost like a highly bureaucratic organization. It has been observed that the manuscript is reviewed several times, sent back to author, and after revision, sent for more revisions. This is very time consuming and surgeons are not able to find time to go over the manuscripts time and again. A reviewer may not agree with the technique described by the author, but should that mean, the manuscript is worthless? There is enormous bias, dogma and prejudice in the medical publishing field. This gets carried over into reviewing and manuscripts are rejected because, sometimes, a reviewer did not think of the idea himself. We wish to eliminate this bias. Anything which is not against ethical practice, not patently harmful will find a place in this journal. In this regard, we are going to follow the principle laid down by Voltaire, "I may not agree with you, but I will defend your right to speak about it". Let the community decide if an idea is worth emulating or not. We will not stop it. We will not censor it. A reviewer or editor is not a censor.

With the academic journal publishing becoming slow and tedious, rejecting case reports and publishing them for a fee, there arise a bunch of predatory journals which will publish anything for a substantial fee. Many surgeons fall prey to such journals and spend huge amounts, hundreds of dollars for publication. This journal will not charge

any publication fee. This is intended to be a diamond open access journal.

We have representations from many countries on the editorial board, namely Afghanistan, Pakistan, India, Nepal, Bangladesh, Vietnam, Cambodia, Malaysia, Indonesia, Singapore, Egypt and England. Many of them are academics with several years experience in teaching and patient care. We hope to provide a judicious mix of academics and practical pediatric surgery in the subsequent issues of the journal.

We aim to support the independent, community based surgeon with lots of enthusiasm and energy, willing to think and innovate, find a working solution for the patient with limited resources. Your contributions, comments, criticisms will only help to improve the journal. Let them come. It takes a year of publishing before the journal can be considered for indexing. The ball is now in your court.

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Editorial

# Transformative Milestones in Pediatric Surgery: Pioneering Advances for Young Lives

Sameh Shehata

*Former President, World Federation of Associations of Pediatric Surgeons (WOFAPS)*

## INTRODUCTION

Pediatric surgery has witnessed remarkable strides in recent years, bringing new hope and improved outcomes for young patients facing complex medical conditions. Surgeons, researchers, and innovators have continually pushed the boundaries of medical science, harnessing cutting-edge technologies and novel approaches to enhance surgical interventions in children. These transformative advances have not only revolutionized pediatric surgery, but they have also brought about profound positive change in the lives of countless young patients and their families.

## MINIMALLY INVASIVE TECHNIQUES: REDUCING TRAUMA AND ENHANCING RECOVERY

Minimally invasive surgery (MIS) techniques have emerged as a game-changer in pediatric surgical practice. By employing smaller incisions and specialized surgical instruments, surgeons can access and treat internal organs with greater precision. This approach has significantly reduced surgical trauma, minimized postoperative pain, shortened hospital stays, and accelerated recovery times for young patients. From laparoscopy to thoracoscopy, MIS techniques have become standard practice for numerous pediatric procedures, including appendicectomy, hernia repair, and many others.

## ROBOT-ASSISTED SURGERY: NEW DIMENSION OF PRECISION

Robot-assisted surgery has opened up new horizons in pediatric surgical care. With the assistance of robotic systems, surgeons can perform intricate procedures with enhanced precision and dexterity. Benefits of this technology include reduced scarring, improved visualization, and improved surgical outcomes. Robot-assisted surgery has proven particularly valuable in complex surgeries such as pediatric urological reconstructions, spinal surgeries, and tumor resections. As this technology continues to evolve, its potential to further optimize surgical techniques and outcomes in pediatric patients is profound.

## FETAL SURGERY: NURTURING LIFE BEFORE BIRTH

Advancements in fetal surgery have expanded the spectrum of possibilities for treating congenital anomalies and genetic disorders even before birth. Fetal surgeries, such as repairing spina bifida or managing diaphragmatic hernia, are performed on the developing fetus within the womb. These interventions, often performed through minimally invasive techniques, can prevent or minimize long-term complications and improve the prognosis for affected children. The remarkable progress in fetal surgery exemplifies the dedication of medical professionals to push the boundaries of science and provide early interventions for the most vulnerable patients.

## REGENERATIVE MEDICINE: UNLOCKING A NEW ERA OF TISSUE ENGINEERING

Regenerative medicine has emerged as a promising field that holds great potential for pediatric surgical care. Stem cell therapies and tissue engineering techniques offer the possibility of regenerating damaged or missing tissues and organs, providing innovative solutions for congenital anomalies and acquired conditions. From skin grafts for burn victims to tissue-engineered bladders, regenerative medicine is paving the way for groundbreaking treatments that restore form and function in pediatric patients. While still in its early stages, the strides made in this field give hope for a future where complex reconstructive surgeries may be replaced by regenerative approaches.

## CONCLUSION

The recent advances in pediatric surgery have brought about a paradigm shift in the way we approach surgical care for young patients. Minimally invasive techniques, robot-assisted surgery, fetal interventions, and regenerative medicine are revolutionizing the field and offering new possibilities for improved outcomes and enhanced quality of life. These remarkable advancements are a testament to the dedication of medical professionals, researchers, and innovators who tirelessly work to push the boundaries of medical science. As we continue to witness transformative milestones in pediatric surgery, we can look forward to a future where young lives are touched by even greater possibilities for healing, recovery, and hope.

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Viewpoint

# In Pursuit of Health Equity for Children with Surgical Needs

Jamshed Akhtar

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Health equity is described as highest attainable health for all people. However, the definition is not uniformly agreed upon.(1) The concept of equity is linked to various factors including political stability, will of the policy makers and prevalence of social justice in general. The United Nations Millennium Development Goals (MDGs) proposed in pursuit of achieving equitable living world for all in year 2000. These were eight in number. The target was to achieve them by 2015. However, same is yet to be attained. It is however good to know that success is reported in accomplishing some of the goals especially those related to communicable diseases, especially the 4-A, that was meant to reduce the mortality in children.(2)

Time moved on and in 2015 another vision was adopted by the United Nations Member States. This was "Transforming Our World: The 2030 Agenda for Sustainable Development." There are 17 Sustainable Development Goals (SDGs). These are interrelated and indivisible as each one is intertwined with other. The vision relates to the prevailing social, economic and environmental challenges. United Nations secretary general Mr. Antonio Guterres in his address has mentioned, "Development is not sustainable if it is not fair and inclusive". It is therefore important to understand the concept of equity specially in the context of healthcare services across the globe irrespective of the gender and age group.(3)

The children in the low and middle income countries (LMIC) suffer the most. Political instability along-with regional armed conflicts for economic gains as well as natural environmental disasters and man-made climate related challenges lead to absolute poverty that deprived people of basic needs including safe drinking water, food, sanitation and many others. Interestingly high birth-rate, a reflection of social needs without considering the responsibility at individual level with limited resources, adds to the perennial challenges in the LMICs. Though healthcare is given a priority in all the policies but same is not visible on ground. The main reasons are the poor governance, corruption and dysfunctional judiciary to name a few.

Surgical conditions of children are not on the priority list of policy makers. With high birth rate, the incidence of congenital anomalies also increases. In addition, trauma, cancers, infectious diseases add to the existing burden. Pediatric patients with surgical diseases do not get a comprehensive care in Pakistan and other developing countries. General pediatric surgeons are over-burdened with huge patient load. In Pakistan, with a population of about 240 million, about 300 pediatric surgeons are registered with the Pakistan Medical-and-Dental Council, the federal licensing body. The number is insufficient even to manage index gastrointestinal malformations like anorectal conditions. Pediatric surgeons are also not

trained to deal with specific surgical conditions like those related head-and-neck (ENT), ophthalmology, plastic-reconstructive surgery, oncology and others. Adult surgeons are not experienced to deal with pediatric and adolescents surgical conditions. There are no fellowship programs in the specialties for comprehensive training for surgical conditions of the children.

The provision of equity gets strengthened when one finds lack of availability of pediatric anesthetists, nurses and rehabilitation services dedicated for the children. Research in the context of needs of the children from LMICs is also lacking. Hardly any funding is available to find out the actual burden of diseases and to set up a registry for congenital anomalies. The recent issue of Seminars in Pediatric Surgery is dedicated to the global surgery (2023Dec).[4] However, the journal charges US dollars 3440 for open access processing for an article. This whooping amount is far from the reach of any clinician and researcher working in the LMICs. In Pakistan, an average pediatric surgeon employed in a governmental hospital gets around US dollars 700 per month as salary. Thus, research is almost non-existent.

National Institute of Child Health in Karachi is the largest tertiary care center in Pakistan for the provinces of Sindh and Baluchistan. It also caters to patients referred from neighboring countries such as the Afghanistan and Iran. More than 40,000 pediatric surgical patients visit outpatient department each year. This number is far from what the institute can handle. There is a long waiting list for elective surgical procedures.

Considering the ground realities and pledges made at different conferences at national and international level, there is growing feeling that voices of those who are at the receiving end are not heard. Most of the agenda remained unattainable as with many of the goals set by WHO and United Nations. Solution to any problem should come from within. The communities themselves should actively take measures to understand their own needs and work on as to how to address them. A true representation from the population is to be identified and workable strategies planned that are applicable to a particular context. At times funding is also available but it is difficult to channelize it in right direction. Social networking and artificial intelligence can help in making strategies with goal oriented time bound tasks.

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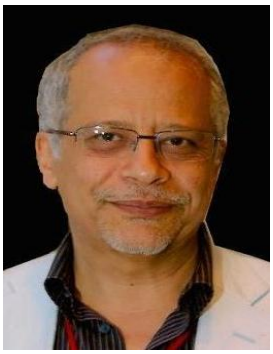
# Welcome to Editors and Editorial Board Members

The *"Pediatric surgery in Tropics"* takes immense pleasure in welcoming the editors and editorial board members. The following is a brief introduction of these accomplished individuals whose achievements easily exceed several pages. Their collaboration is sure to enrich the exchange of knowledge among pediatric surgeons around the globe.



**Sameh Mahmoud Shehata** (Patron)

Dr Shehata is Professor and Past-Chairman of the Department of Pediatric Surgery at the Faculty of Medicine, University of Alexandria, Egypt. He was President of the Egyptian Association of Pediatric Surgeons (EPSA), IPEG Middle East Chapter and President of the World Federation of Associations of Pediatric Surgery (WOFAPS). He innovate a technique of laparoscopic orchidopexy (Shehata technique). He is editorial board of many national and international pediatric surgery journals.

**Vivek Gharpure** (Editor-in-Chief)

Dr. Vivek Gharpure is Professor of Pediatric Surgery at the Rural Medical College, Ahmednagar, India. Formerly he was Associate Professor at MGM Medical College and Government Medical College, Aurangabad. He was Vice President of Maharashtra chapter of Indian Association of Pediatric Surgeons and Convener of Community Oriented Pediatric Surgeons (COPS). He is an avid birdwatcher, marathoner, mountaineer, bibliophile, origami artist and he likes to make mathematical puzzles. He volunteers for two weeks every year to work for charity in pediatric hospitals of South East Asia.

**Venkatachalam Raveenthiran** (Editor)

Dr. Raveenthiran is Professor and Head of Pediatric Surgery at Government Cuddalore Medical College, Chidambaram, India. Formerly, he was Professor and Head of Pediatric Surgery at Annamalai University and SRM University. He was Associate Editor of the *Indian Journal of Surgery*, Editor of the *Journal of Neonatal Surgery,* Editor-in-Chief of the *Journal of International Medical Sciences Academy and* Editor of *Yakkai*, a vernacular (Tamil) Health Magazine. He is an editor of *IAPS Textbook of Pediatric Surgery*. He has authored 120 journal articles and 14 chapters in textbooks with an h-index of 22. His articles are cited in more than 99 popular textbooks. He was President of the Association of Tamilnadu and Pondicherry Pediatric Surgeons and Founder Convener of Community Oriented Pediatric Surgeons. He is a Tamil poet and has authored 3 literary books in Tamil. His name is eponymously associated with ectopic testis within Spigelian hernia (Raveenthiran Syndrome).

**Yogesh Kumar Sarin** (Editor)

Dr. Yogesh Kumar Sarin is Director Professor and Head of the Department of Pediatric Surgery at Lady Hardinge Medical College, New Delhi. Prevously he served as Head of Pediatric Surgery at Maulana Azad Medical College, Delhi. He is also qualified MBA in health care. He is recipient of many awards and honors including Delhi State Award for Doctors for the year 2012-13. He is a visiting Professor at many universities/ teaching hospitals in India and abroad. He has to his credited 300 publications with an h-index of 19**.** He authored a monograph on Wilms' Tumor, besides several book chapters. He is editor-in-chief of the *Journal of Neonatal Surgery* and editorial board member of more than a dozen journals. He has been a reviewer for 76 medical journals. He was the Secretary-cum treasurer (2005-07) and President of Indian Association of Pediatric Surgeons (2021-22). He is presently Secretary of Indian Child Abuse, Neglect and Child Labor (ICANCL) group of Indian Academy of Pediatrics. He is the master mind behind the *Annual Updates in Pediatric Surgery* at Maulana Azad Medical College which is very popular among Pediatric Surgical trainees.

**Govindarajan Krishnakumar** (Assistant Editor)

Dr. Krishnakumar is Professor of Pediatric Surgery at the Jawaharlal Institute of Postgraduate Medical Education and Research, Pondicherry, India. He is a Fellow of several prestigious organizations including the American College of Surgeons, International College of Surgeons, Association of Surgeons of India, Indian Association of GI Endoscopic Surgeons, Indian Association of Pediatric Surgeons and Indian Society of Pediatric Urology. He has published more than 50 publications, 5 book chapters. He is reviewer for several prestigious journals.

**Ananda Kumara Lamahewage** (Editorial Board Member)

Dr Lamahewage is Senior Consultant Paediatric Surgeon at Lady Ridgeway Hospital, Colombo, Sri Lanka. He is President of the Sri Lankan Association of Pediatric Surgeons. He is a member of World Federation of Associations of Pediatric surgeons, SAARC Federation of Pediatric surgeons and College of Pediatricians of Sri Lanka. He is a Founder member of the Global Initiative for Children's Surgery.

**Ashish Lal Shrestha** (Editorial Board Member)

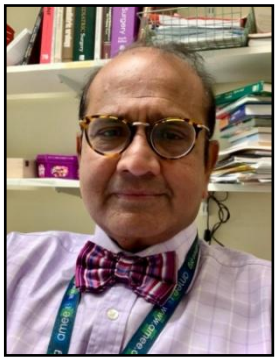
Dr. Ashish Lal Shrestha is Associate Professor and Head of the Department of Pediatric and Neonatology, Kathmandu Medical College and Teaching Hospital, Kathmandu, Nepal. He is Consultant Pediatric and Neonatal Surgeon, Grande International Hospital, Kathmandu.

**Humberto Lugo Vincente** (Editorial Board Member)

Dr Humberto Lugo Vincente is Professor Pediatric Surgery and Academic Director of Pediatric Surgery at Department of Surgery, Hospital Regional de Bayamon at Universidad Central del Caribe, School of Medicine at University of Puerto Rico and Ponce School of Medicine, Puerto Rico. He is also Consultant Pediatric Surgeon at San Jorge Children's Hospital. He is Editor-in-Chief of *Pediatric Surgery Update, Pediatric Surgery Handbook* and *Boletin Association Medicine PR.* He was Chairman of American College of Surgeons (Puerto Rico Chapter). He is editorial board member of several journals. He was Guest Editor of *Seminars in Pediatric Surgery*. He is the Founding Member "Zonamd.com". He has authored 56 research articles and several book chapters. He is reviewer of many leading pediatric journals. He is recipient of several awards including Doctor of the Year – Centro Medico San Pablo 1995. He was the first to introduce pediatric laparoscopy in Puerto Rico. Painting is his hobby and he has conducted several Oil-Painting Expositions.

**Jamshed Akhtar** (Editorial Board Member)

Dr Jamshed Akhtar is Visiting Professor at Department of Pediatric Surgery, National Institute of Child Health, Karachi, Pakistan. Formerly, he was Dean, Faculty of Pediatric Surgery at the College of Physicians and Surgeons of Pakistan. He was President of Pakistan Association of Medical Editors (PAME) and is a Visiting Faculty at the University of Health Sciences and Shalamar Medical & Dental College, Lahore. He was Editor of the Journal of College of Physicians & Surgeons Pakistan (JCPSP). He is member of the National Bioethics Committee, Government of Pakistan. He is the Course Director of ATLS program and is lead instructor BLS and PALS courses of American Heart Association. He is member of World Association of Medical Editors and Eastern Association of Medical Editors



**Varadarajan Kalidasan** (Editorial Board Member)

Dr. Kalidasan is Honorary Consultant Pediatric Surgeon cum Urologist at University Hospitals Sussex. He is Governor in Board of Governor of NHS foundation trust, Brighton, United Kingdom. He is also Honorary Clinical Education Tutor of the Royal College of Surgeons, Edinburgh. He is a Member of International Affairs Committee of the British Association of Pediatric Surgeons. He is the Chair of Southeast division of British Association of Physicians of Indian Origin. His interest and eminence in Cricket game earned him the post of Director, Board of Sussex Cricket Limited.

**Mohamed A Baky Fahmy** (Editorial Board Member)

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**Naeem Khan** (Editorial Board Member)

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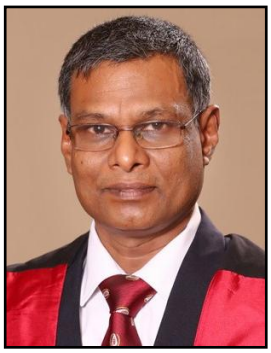

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Case Report

# Pristine Cystine Urolithiasis

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**Keywords**
Cystine calculus
Urolithiasis
Cystinuria

**Abstract**

*Urolithiasis in children is rare with a reported incidence of 1.8 per 1000 children. Cystine calculi are still rarer with a prevalence of 1:7000 and the typical age of onset is the second decade of life. We present a 3 years old girl with recurrent cystine stones in kidney.*

## INTRODUCTION

Cystine was described as an unusual chemical component in bladder calculi by Wollaston[1] about two centuries ago. He termed it cystic oxide. This nomenclature was later changed by Berzelius [2] to cystine because of the absence of an oxide component in it. The chemical structure of cystine was elaborated by Friedman[3] in the early 20th century. Garrod hinted as early as 1908 that cystinuria might be caused by an inherited disorder.[4]

The autosomal recessive pattern of cystinuria was first accurately described by Harris in 1955.[5] Its primary manifestation is repeated stone formation. Prevalence of cystine calculi is 1:7000 and the typical age of onset is in the second decade of life. [6,7] Urolithiasis per se is rare in children with reported incidence of 1.8 per 1000 children.[8] Cystine stones constitute 1–2% of all urinary calculi and 6–8% of pediatric renal calculi.[2] High recurrence rate of 60% with the accompanying risk of progressive renal impairment[9] in cystine urolithiasis justifies regular follow-up examination of patients with cystinuria.

We report a 3 years old girl with recurrent cystine calculi of the kidney and bladder. To our knowledge pure cystine renal calculi in children has rarely been reported in Indian medical literature.

## CASE REPORT

A 3-year-old girl presented with abdominal pain and difficulty during micturition for 6 months. She was diagnosed with urinary bladder calculus along with left renal calculus. Patient underwent open cystolithotomy followed by left open pyelolithotomy after 3 weeks. After one year interval, she presented with similar complaints, investigation revealed left sided staghorn calculi causing pelvic-ureteric junction obstruction. She again underwent left pyelolithotomy. Postoperative recovery was uneventful.

Renal stone analysis showed 100% cystine stone. (Fig 1) Urine was found to be highly acidic with a

pH ranging from 3 to 4. Twenty-four hours urine analysis did not show cystine crystals. X-ray diffraction of the renal stone showed presence of cystine. (Fig 2)

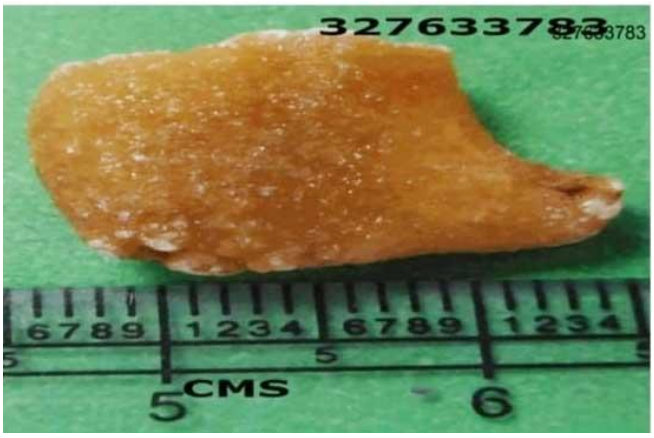


**Fig 1.** *The cystine stone.*

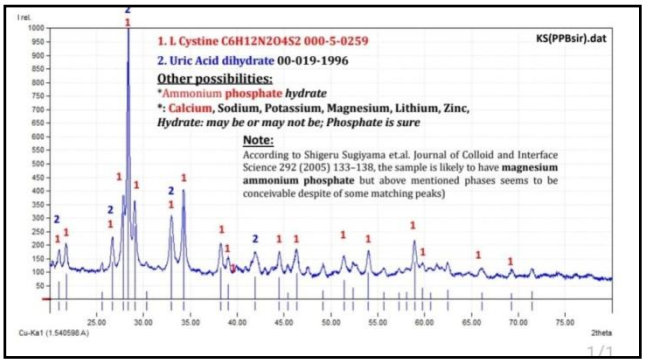


**Fig 2.** *Report of X ray diffraction analysis of the stone*

## DISCUSSION

Renal stones may form at any age; more than 80% of patients develop their first stone within the first 2 decades.[10,11] Early formation of stones is more likely in males than in female patients. Our patient presented with cystine stone at a young age of 3 years.

Cystine is poorly soluble at physiological urine pH between 5 and 7. Stones are formed especially when urinary cystine levels exceeds 240–300 mg/l (1.33–1.66 mmol/l). Higher pH value of >8 leads to threefold increase in solubility of cystine crystals thus preventing stone formation.[12] Consequently, urine alkalinization is the main goal of pharmacotherapy. Oral alkalinization treatment is a safe and effective way of keeping the urine pH between 7.5 and 8.0. Potassium citrate is safer than the sodium compound (starting at 60–80 mEq/day).[13] In our patient, urinary pH was found to be between 4 and 5; after citrate supplement it was 8. Cystine stones must be suspected in patients, with family history of cystinuria.[14] In our patient the family history of cystinuria or recurrent urolithiasis was absent.

Microscopic examination of the first voided urine may reveal typical hexagonal cystine crystals, that confirm the diagnosis. However, such crystals are detectable in only 20–25% of urine specimens in patients with cystinuria.[11,13] Cystine stones typically have a homogeneous structure without striations and are visualized easily on plain radiographs, but they are less radio-dense than struvite or calcium oxalate stones.[15] Stag-horn cystine calculi are common.

Primary goal of conservative treatment is (in children and adults) to increase the solubility of urinary cystine. It includes increased fluid intake, low-salt diet and urinary alkalinization. Maintaining a high urine output of more than 120 ml/h is essential for therapeutic success regardless of drug treatment.

Compliance of these patients with medical treatment is often poor and most of them experience recurrent episodes of stone formation, requiring multiple interventions. Frequent ultra-sonography should be done to identify early recurrence. Family members of patients with cystinuria should undergo screening.[16,17]

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Clinical Research

# Factors Influencing the Survival of Patients with Gastroschisis in A Lower-Middle Income Country: A Retrospective Observational Study

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**Keywords**
Gastroschisis
Abdominal wall defects
Silo Treatment

**Abbreviations**
**LMIC** - Low and middle income countries

## Abstract

***Background:*** *Survival rate of gastroschisis has improved worldwide due to advancements in antenatal diagnosis, perinatal care and neonatal intensive care. This is in stark contrast with the persistently poor outcomes observed in low and middle-income countries. Aim of this study is to identify factors affecting survival in a resource constraint center, where intensive neonatal care and parenteral nutrition could not be provided.*

***Methods:*** *This single-center retrospective observational study was done at our institute from September 2022 to 2023. Data were collected from hospital records. All patients admitted with gastroschisis during the study period were included and grouped into A (survivor) and B (non-survivor). Factors influencing the outcome were compared between the two groups. All patients were systematically categorized based on antenatal diagnosis, age at presentation, maturity, birth- weight and mode of repair. Statistical analysis was done using SPSS version 22.*

***Results:*** *Among a cohort of 48 patients, only 8 (16%) survived. Twenty patients presented within 24 hours of birth of whom 5 survived. Age at presentation did not significantly impact the outcomes (P=0.25). Seven patients were full term and no statistically significant difference in terms of maturity was observed between the two groups (P=0.05). A pre-formed silo procedure done in 30 patients yielded a survival rate of 13.3% with four individuals demonstrating positive outcomes. Twelve patients underwent traditional silo procedures and only one of them survived. Among the 6 patients who underwent primary repair, 3 died and 3 survived.*

***Conclusion****: Without adequate intensive care and nutritional support, no technique of closure can improve the survival rate of patients with gastroschisis. Prompt antenatal diagnosis and enhanced perinatal care are critically important to improve the outcome of gastroschisis in resource-constrained settings.*

## INTRODUCTION

Gastroschisis is a congenital defect of anterior abdominal wall on the right side of umbilical cord causing protrusion of abdominal contents through the defect without any covering of amniotic membrane.[1] Incidence of gastroschisis is 1 in 4000 live birth.[2,3] The mortality rate ranges between 33 and 100% in the low and middle-income countries (LMIC). In contrast, survival rate is above 95% in high income countries with the advanced antenatal diagnosis, improved perinatal care and intensive neonatal care.[4,5]

Neonates with gastroschisis are at the risk of losing water and body heat from the exposed bowel, compromised intestinal circulation, and mechanical irritation to the gut.[5] Factors known to influence the survival of gastroschisis are antenatal diagnosis, time of delivery, gestational age, birth weight, time lapse in reaching the tertiary care center, associated anomalies and the surgical technique.[6,7] In high income countries, uncomplicated gastroschisis has very low morbidity with a mortality rate nearing zero. But in developing countries like Bangladesh, mortality rate is still high.[6] In LMIC accessing specialist care is delayed due to absence of prenatal recognition and poor neonatal referral and transport system.[9] When there is delay in initial management, the exteriorized gut becomes matted and edematous, which makes its replacement into abdominal cavity difficult.[10]

Depending on the accommodation capacity of abdomen and the condition of the gut, either primary fascial closure or staged closure (using traditional hand sewn silo or spring loaded preformed silo) is performed to reduce the exteriorized gut and close the defect.[11] In comparison to delayed closure, primary repair of gastroschisis is associated with improved neonatal outcomes, including early initiation of enteral feeds, early discontinuation of parenteral nutrition, shorter hospital stay and lower risk of surgical wound infection.[12] When delayed closure is resorted to, several studies have shown the benefits of preformed silo in low resource center as it is thought to avoid abdominal compartment syndrome thereby circumvents the need of ventilatory support.[9] However, staged-closure demands longer period of parenteral nutrition as enteral feeding cannot be started until fascial closure is achieved.[13]

Aim of this study is to identify the factors contributing to the survival of neonates with gastroschisis, where neonatal intensive care and parenteral nutrition could not be provided.

## MATERIALS AND METHODS

This single-center retrospective observational study was done at Bangladesh Shishu Hospital and Institute from September 2022 to 2023. All new born with gastroschisis admitted to this hospital were included. Neonates with gastroschisis, who were discharged against medical advice, were excluded from the study. Clinical data retrieved from hospital records included age, sex, antenatal and postnatal history, gestational age, birth weight, mode of delivery, clinical findings, time from delivery to hospital admission, and associated anomalies. Management was initiated with resuscitation and stabilization of the neonate by maintaining a warm environment, keeping the baby dry, and preventing heat loss. Immediately after admission, herniated viscera were kept enclosed in a sterile saline-bag. Nasogastric decompression and bladder catheterization were done. Intravenous access was established for fluid resuscitation and broad-spectrum prophylactic antibiotics were started. The choice of surgical procedure (primary closure *vs.* silo application) was decided according to the general condition of the neonate, the size of defect, nature of herniated organs and presence of viscera-abdominal disproportion (VAD). Post-operatively, patients were kept on nothing per oral and nasogastric suction. In case of silo repair, it was squeezed every day and final repair was performed when complete repositioning was possible.

Clinical data were carefully logged in the Excel sheet. All patients were grouped into A (survivor) and B (non-survivor). Factors influencing the outcome were compared between the two groups. The Statistical analysis was done by using a statistical package for social sciences (*SPSS*). Continuous variables were compared with student t-test and discrete variables were analyzed with chi-square test where applicable. A p <0.05 was considered as significant.

## RESULTS

A total of 50 neonates with gastroschisis were admitted during the study period. Two neonates were excluded as they were discharged against medical advice and the remaining 48 were included in this study. Among these 31 were male and 17 were female. Only 8 (16%) of them survived. Prenatal diagnosis was made in only 3 patients, among them two (66%) patients survived.

Table 1: Factors affecting the survival of gastroschisis

| **Factors** | **Group A** n=8 | **Group B** n=40 | ***P*** |
|---|---|---|---|
| **Antenatal diagnosis** (n=3 | 2 | 1 | - |
| **Age at presentation** | | | |
| < 24 hr (n=20) | 5 | 15 | 0.25 |
| > 24 hr (n=28) | 3 | 25 | |
| **Maturity** | | | |
| Term (n=25) | 7 | 18 | 0.05 |
| Preterm (n=23) | 1 | 22 | |
| **Birth Weight in Kg** | | | |
| (Mean ± SD) | 2.32 (± 0.28) | 2.13 (± 0.35) | 0.56 |
| **Mode of repair** | | | |
| Preformed silo (n=30) | 4 | 26 | 0.06 |
| Traditional silo (n=15) | 1 | 11 | |
| Primary repair (n=6) | 3 | 3 | |

A total of 20 patients (42%) were admitted to the hospital within 24 hours of birth; among them 5 (25%) survived. The remaining 28 patients (58%) reached hospital after more than 24 hours of birth; among them 3 survived (10%). There were 23 preterm (48%) and 25 (52%) term neonates (Table 1). Among the preterm neonates, only one survived (4%). However, no significant difference ($P = 0.05$) was seen in terms of maturity in the survivor and non-survivor groups. Mean birth weight was 2.32±0.28 kg in the Group A and 2.13±0.35 kg in the Group B. The small intestines were herniated 100%, the colon in 83%, and the stomach in 60% of the patients. Pre-formed silo application was performed in 30 patients, of which 4 survived. Traditional silo was performed on 12 patients, of which 1 survived. Among the 6 patients who underwent primary repair, 3 survived (Table 1).

Death occurred within 48 hr of hospital admission in 24 patients (60%) and the causes of death in these patients were late presentation (>24 hours of birth), hypovolemia, hypothermia, and sepsis. Among the primary closure group, 2 patients expired within the first 72 hours due to abdominal compartment syndrome, multi-organ failure, and sepsis. In our patients, the diagnosis of abdominal compartment syndrome was made based on clinical signs and symptoms such as abdominal distension, paralytic ileus and oliguria. Duration of hospital-stay in gastroschisis survivors ranged from 1 to 4 weeks.

## DISCUSSION

The incidence of gastroschisis has risen through-out the world in the last 3 decades.[14–16] A survey was carried out among pediatric surgeons by Wright and colleagues in 2012. They reported 2 cases on an average per institution per year in low-income countries, while middle-income and high-income countries reported 12 and 15 cases per institution per year, respectively.[17] A study was performed in our institution in 2017 found 75 cases within 3 years.[6] In the present study, we

found 48 cases within one year. In the absence of formal registry for congenital anomalies in Bangladesh, it is difficult to calculate the exact incidence.

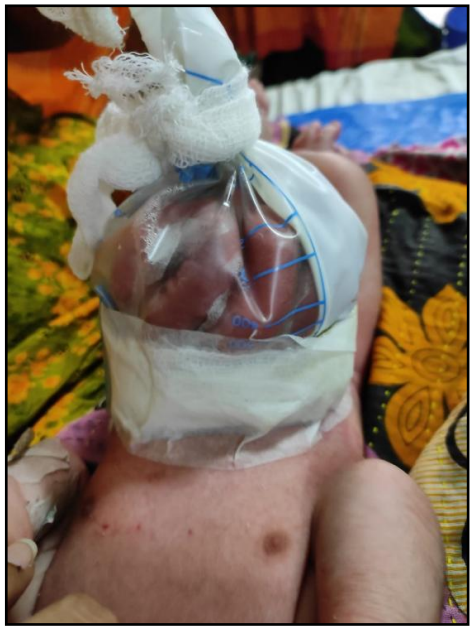

**Fig 1.** *Traditional silo*

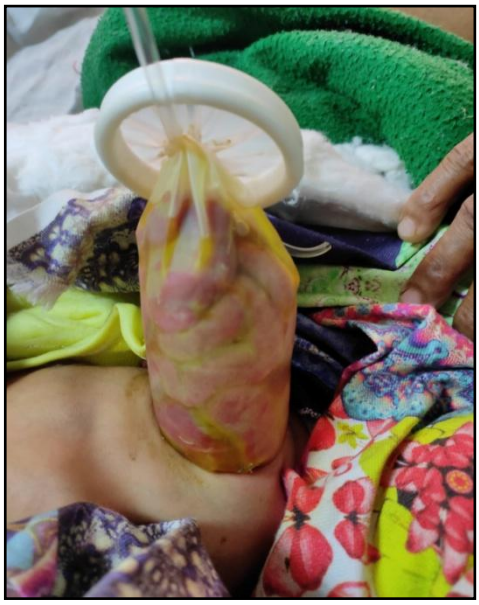

**Fig 2.** *Preformed silo*

Birth weight is an important factor affecting the outcomes. In a study, no newborn with a birth weight less than 1500 gm survived.[1] We also observed that survival was more in term neonates with higher birth weight, though the difference was not statistically significant. In developed countries, most of the gastroschisis are diagnosed antenatally and these neonates are delivered in a specialist center where surgery can be performed. In contrast to the high income countries, these defects are diagnosed postnatally at primary health care centers in Bangladesh and then referred to tertiary referral institutes. In our study, 20 mothers had antenatal ultrasound scan at least once during pregnancy. But only 3 had the diagnosis; they all had their delivery at a private hospital from where the neonates were shifted to our hospital within 24 hours of birth and 2 of them survived. We have observed that antenatal sonographers often focus on basic obstetrics parameters and fetal viability rather than detection of congenital anomalies. Similar observation was made by Gurjar.[1] In their series of 58 cases, no one had an antenatal diagnosis. In our study, most of the neonates were delivered at home, primary health-care clinics or secondary health-care centers, and these babies were transported, usually over long distances, without adequate initial resuscitation. Most of these patients arrived at our hospital without a functional intravenous line or any protective covering of the exposed gut. A study by Stevens *et al* showed that poor resuscitation is a more significant predictor of mortality than postnatal transfer time. They highlighted the importance of adequate initial care.[19] Likewise, the survival rate was less in our study in those who presented after 24 hours of birth. Among the 48 cases, 28 reached our hospital after more than 24 hours of birth and 25 (89%) of them expired.

The intestinal morbidity observed in patients with gastroschisis may be due to the damage of the bowel wall that occurs by exposure to amniotic fluid as well as by constriction of the intestine and its blood supply at the abdominal defect.[20–22] Based on this hypothesis, elective preterm delivery by caesarian sections is the standard management in some parts of the world.[23,24] On the contrary, some authors mentioned that there is no advantage of routine preterm delivery.[18]

Baud *et al* found induction at 37 weeks was associated with a lower rate of sepsis, bowel damage, and neonatal death compared with pregnancies managed expectantly beyond 37 weeks.[25] In our study term neonates survived more than preterm neonates, though the difference was statistically not significant. It supports the hypothesis that there is no survival advantage of routine preterm elective delivery in gastroschisis.

The abdominal wall closure method is thought be an influencing factor in the outcome of gastroschisis. Fraga *et al* found that primary closure has benefits over delayed closure.[18] In our study, we did not find any significant difference in the outcomes following different approaches of reduction. In fact, primary reduction is possible only when the intestine is in good condition (thin wall, no peel) and abdominal cavity is adequate. Patients who have the intestine in worse condition (wall edema, thick peel, shortened mesentery) generally undergo silo reduction. Exposed viscera in almost all of our cases (42 out of 48) were swollen and edematous. These patients were in severe hypovolemia and hypothermia. Thus, silo (preformed or traditional) was performed in most patients though primary repair is the treatment of choice. We used traditional silo only when it was difficult to accommodate the contents within the preformed silo bag or when the preformed silo was unavailable. The survival rate did not differ significantly and hence we could not conclude that one closure method was better than the other. However, several studies have shown that the use of preformed silos is associated with lower requirements for mechanical ventilation, reduced time to feeding, lower infection rates and lower risk of abdominal compartment syndrome.[13,26,27]

In our study, 60% of our patients died within 48 hours after hospital admission. Most of them suffer from hypothermia, sepsis, and acidosis. Our institution, like other developing countries, has significant difficulty in caring for these patients in an appropriate ICU setting due to a lack of availability of NICU beds. Non-availability of total parenteral nutrition was also a challenge in our country in managing these babies. On the contrary, parenteral nutrition, and neonatal intensive care support were invariably given in developed countries where the survival rate of gastroschisis was more than 95%.[9]

This present study is limited by the relatively small sample size from a single center and by the retrospective nature of the research. The survivor group was too small to reflect any statistically meaningful conclusion. Another limitation is long-term outcome could not be evaluated due to poor follow-up and resource limitations

To conclude, inadequate antenatal diagnosis and delay in initial resuscitation are the important factors that contribute to poorer outcome of gastroschisis. Condition of the bowel, adequate nutritional and intensive care support rather than the operative technique appears to determine favorable outcome.

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Randomized Controlled Trial

# Fast-Track Protocol *versus* Conventional Protocol for Colostomy Closure in Children: A Randomized Controlled Trial

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**Keywords**
Colostomy

**Abbreviations**
**CP** - Conventional protocol
**ERAS** - Enhanced recovery after surgery
**FTP** - Fast-track protocol

## Abstract

***Background:*** *Recent literature has focused on Enhanced Recovery After Surgery (ERAS) Programs or Fast Track Elements in peri-operative care of patients undergoing surgery. While fast-track surgery has been studied extensively in adult patients, studies in the pediatric age group are rather few. Existing few studies are disadvantaged by non-homogenicity of surgical procedures. For colonic surgeries, only early postoperative enteral feeding has been studied in detail. There are no randomized controlled trials that evaluate fast-track protocol in colonic surgery of children.*

***Methods:*** *Thirty children with colostomy for different indications were randomized into two groups of 15 each. The control group had conventional protocol (CP) which included antegrade intestinal irrigation, conventional preoperative enemas postoperative nasogastric suction, placement of abdominal drains and slow commencement of oral feeds. The trial group followed the Fast Track protocol (FTP) that included parent and patient education, no bowel preparation, no prolonged pre-operative fasting, avoidance of routine placement of drains, tubes and catheters, immediate post-operative feeding, epidural anesthesia, immediate mobilization and limiting systemic opioids. The outcomes of two groups were compared.*

***Results:*** *FTP leads to a significant decrease in post-operative pain, time taken to sit-up and stand, incidence of the overall complications and wound infection. However, it is associated with a significant increase in the rate of ileus. There is no significant difference between the two protocols as regard to the overall durations of hospital stay, return of bowel function, catheterization rate, readmission rate and time taken to walk and play.*

***Conclusion****: Our study suggests that the conventional practice of bowel preparation, prolonged fasting, insertion of nasogastric tube and drain, systemic analgesia, delayed feeding, and post-operative bed rest are not mandatory in children undergoing colostomy closure. Fast-track protocol appears to be safe and effective.*

## INTRODUCTION

Traditionally, colostomy closure involves mechanical bowel preparation with antegrade intestinal irrigation and conventional enemas.[1] Patients are kept fasting for long hours pre-operatively to prepare the bowel[2] as well as to decrease anesthesia-related aspiration pneumonitis.[3] Intra-operative drain placement below the fascia was advocated in the past.[2] All patients are routinely placed on nasogastric tube drainage with intravenous hydration. Oral feeds are begun slowly as the gastric dysmotility resolves.[4] Post-operative analgesia mainly consists of systemic (intravenous or oral) analgesia with opioids and non-steroidal anti-inflammatory drugs. All these measures are a hindrance for early ambulation that is prevented by multiple tubings attached to the patient.

Recent literature has focused on the use of *Enhanced Recovery Programs* or *Fast-Track Elements* in peri-operative care of the patients undergoing surgery. They include parent and patient education, no bowel preparation, no prolonged pre-operative fasting, minimally invasive surgical techniques, avoidance of routine placement of drains, tubes and catheters, immediate post-operative feeding, immediate mobilization as well as multimodal analgesia including loco-regional analgesia where feasible and limiting systemic opioid drugs.[5-7]

While fast-track surgery has been studied extensively in adult patients, studies in the pediatric age group are very few. Most of the in pediatric studies include a mixed variety of surgeries. For colonic surgeries, only early postoperative enteral feeding has been studied in detail. There have been no randomized controlled trials to evaluate fast-track colonic surgery in pediatric patients.

We aimed to assess the outcomes using a FTP for peri-operative management for colostomy closure in children and compare them with that of the conventional protocol (CP). We attempted to find out if the use of FTP leads to a decrease in hospital stay and other benefits such as reduced post-operative pain, early return of bowel and bladder function, ambulation and reduced morbidity.

## MATERIAL AND METHODS

The study was conducted in a tertiary care centre of Pediatric Surgery in 2013-14. All children below 12 years of age and American Society of Anesthesiology class I and II, who are undergoing colostomy closure, were evaluated. Exclusion criteria included contraindications to neuroaxial anaesthesia (e.g. gross spinal anomaly, infection, or skin lesions over the back), distal loopogram showing disuse atrophy of colon in children having colostomy for more than 6 months and revision/redo of colostomy done for any cause. Those patients who met the necessary inclusion criteria were recruited in the study after institutional ethics committee clearance and the informed consent of parents and assent from children aged more than 7 years.

After going through a similar study[5] in adults done by Nanavati *et al*, the sample size was estimated to be four in either group to achieve a confidence level of 95% and a power of 80%. Sample size was calculated using the formula: $(1.645+0.84)^2$ X (sum of squares of standard deviation) / (square of the difference between the two means). However, we decided to recruit 15 patients in each group. Thirty such patients were chosen and were randomly divided into two groups using computer software (simple randomization) – 15 patients were managed using the conventional protocol (CP) and 15 patients were managed with fast-track protocol (FTP).

On the day of operation, all the patients (both groups) were taken-up as the first case in the morning and given general anesthesia. Fentanyl was given intravenously. Propofol, Isoflurane in Nitrous oxide and Oxygen and Vecuronium were used for anesthesia. The anastomosis was done with simple interrupted sutures (single layer) using polyglactin suture material.

The controls (CP group) were managed as per the conventional protocol for stoma closure that was being followed in our unit. Preoperative measures included mechanical bowel preparation with distal stoma washes and polyethylene glycol solution (1g/kg) with a glass of water given 16 hours before surgery. Solid diet and breast milk were restricted 16 hours before surgery; clear liquids were allowed up to 6 hours before surgery. Intraoperatively, a nasogastric tube was routinely placed and an intra-abdominal corrugated drain was placed. Post-operative measures included pain relief with opioids (Tramadol 1mg/kg/dose intravenously 8 hourly) and non-steroidal anti-inflammatory drugs (Paracetamol 15mg/kg/dose intravenously or orally three times a day). Ketorolac was used as a rescue analgesic (when pain score >6 despite administering the above mentioned analgesia), given intravenously (0.4mg/kg) SOS up to a maximum of three doses in a day. The nasogastric tube was removed after the return of bowel sounds and when the nasogastric aspirate turned non-bilious. A liquid diet was begun after the removal of the nasogastric tube. The drain was removed once soakage was less than one gauze piece in 24 hours. Intravenous fluids were given when the child was in nil by mouth.

Fast-track elements that were implemented in the cases (FTP group) included:

1. Pre-operative counseling of the parents and children.
2. Avoidance of mechanical bowel preparation.
3. Preoperative fasting of six hours for solids, four hours for breast milk and two hours for clear liquids.
4. Avoidance of prophylactic placement of nasogastric tube and abdominal drain.
5. Placement of an epidural catheter at L2-3 level (for sigmoid colostomy) or T9-10 level (for transverse colostomy). At the start of surgery, 0.25% bupivacaine was given. At the end of surgery, 30 μg/kg of morphine in 2-6ml of saline was given epiduraly. Adequate pain relief in the form of epidural morphine (30 μg/kg in 2-6ml of saline once a day) was given. Epidural catheter was kept *in situ* for 2 days after surgery. The patient was also given oral non-steroidal anti-inflammatory drugs (intravenous or oral syrup Paracetamol 15mg/kg three times a day). The rescue analgesia protocol was the same as was for the controls (CP group)
6. Early mobilization of patients in the post-operative period. Children were encouraged to sit-up on the evening of surgery and walk on postoperative day-1. They should be ambulated at least 4 times for 10 minutes each time on postoperative day-1. Parents of infants were encouraged to hold the baby upright for half an hour at least once on the evening of the day of surgery itself and at least 4 times on postoperative day-1.
7. Early enteral feeding within the first 24hr is started with sips and clear liquid diet on post-operative day-1 and solids on post-operative day-2. Exclusively breastfed infants fed on postoperative day-1. Intravenous fluids were given while the child was on nil-by-mouth.
8. Prevention of postoperative nausea and vomiting with antiemetic drugs (Ondansetron 0.1 /kg intravenously tds - max 4mg/day)

In case, there was any suspicion of an anastomotic leak or obstruction based on the clinical or hematological parameters, an ultrasound and X-ray abdomen (erect) were done. If the suspicion was confirmed, then the fast-track protocol was abandoned.

The criteria for discharge were the same for both groups: i.e. accepting a full diet, having passed flatus or feces and being able to carry out daily routine physical activity as per the age. All the patients were evaluated during their hospital-stay and were called for follow-up one week after discharge.

The primary outcome evaluated in patients in both groups was duration of hospital stay. The secondary outcomes included:

1. Pain on the evening of the days of surgery (at 9pm) and on the post-operative days 1 and 2 (at 9am). It was assessed using the FLACC Behavioral Pain Assessment Scale for children less than 4 years of age and the FACES Pain Scale for children 4 years and older.
2. Return of bowel function as assessed by auscultation of bowel sounds, asking for passage of flatus and/or feces and acceptance of diet.
3. Time at which child first voids urine spontaneously after surgery.
4. Readmission to the hospital, and the underlying reason(s).
5. Time at which the child returns to school or engages in playful activities (in children >1 yr).
6. Post-operative complications like vomiting, wound infection, wound dehiscence, paralytic ileus, anastomotic leak, obstruction, and itching (due to epidural analgesia)

Results between the two groups were compared by protocol–based analysis to see if there was any statistically significant difference in the above parameters. The quantitative variables were expressed as mean ± standard deviation (sd) and compared using the unpaired t-test/Mann-Whitney test. Qualitative variables were expressed as frequencies or percentages and compared using Chi-square/Fisher's exact test. Statistical Package for Social Sciences (SPSS) version-15.0 software was used for statistical analysis. Statistical significance was set at P <0 .05

## RESULTS

The indications of colostomy in these 30 children included anorectal malformation (n=23), perineal injury (n=4) and bowel perforation (n=3). The cases and controls were age- and gender-matched with no statistical differences. The sigmoid colostomy was the commonest procedure followed by the transverse colostomy. Divided colostomy was slightly more common than the loop colostomy.

The primary duration of hospital stay (duration of hospitalization after first admission) ($P$= 0.282) and overall duration of hospital stay (including readmissions) ($P$= 0.119) did not differ significantly in the two groups. The pain scores on the day of surgery and post-operative days 1 and 2 were significantly lower for the FTP group as compared to the CP group (Table 1). The number of doses of rescue analgesia (ketorolac) required for the FT group was significantly lower than that needed for the CV group (Table 2).

Table 1: Pain Scores

| Pain Score | Day 0 | Day 1 | Day 2 |
|---|---|---|---|
| **CP** Mean ± SD | 5.33 + 1.50 | 4.40 + 0.91 | 4.47 + 0.92 |
| **FTP** Mean + SD | 3.60 + 2.13 | 2.93 + 1.71 | 2.93 + 1.10 |
| **p-value** | 0.008* | 0.003* | <0.001* |

**Statistically significant (using unpaired t-test)*

Table 2: Number of Rescue Analgesic (Ketorolac) Doses Required

| Number of rescue analgesic | CP (n=15) | | FTP (n=15) | |
|---|---|---|---|---|
| | n | % | n | % |
| 0 | 9 | 69 | 14 | 93 |
| 1 | 3 | 23 | 1 | 7 |
| 2 | 1 | 8 | 0 | 0 |
| Mean ± SD | 0.38 ± 0.65 | | 0.07 ± 0.26 | |

*p-value 0.046 (Statistically significant; Mann-Whitney test); CP - Conventional protocol; FTP - Fast-track protocol*

Return of bowel function was earlier in the FTP than the CP group; but the difference was not statistically significant. Young children could not express passage of flatus and thus could only be recorded in 10 cases each group. Infants were on exclusive milk feeds; thus, acceptance of a solid diet could only be measured in 11 patients of the CP group and 13 of the FTP group.

In the CP group, the average times to removal of the nasogastric tube and the abdominal drain

were 49.87 (± 21.96) hrs and 2.53 (± 0.74) days respectively. None of the children was routinely catheterized intra-operatively. However, 1 patient in the CP group and 2 patients in the FTP group went into urinary retention and required catheterization post-operatively; catheters were removed on post-operative day-2.

Two of the patients in the CP group required readmission. One patient developed a fecal fistula that resolved on conservative management. This child required readmission for six days. The other child required readmission (3 days) for a wound infection that was managed with intravenous antibiotics and dressing.

The time taken to sit-up and stand was significantly lower in the FTP group as compared to the CP group.(Table 3) As some of the infants were too young to have physical mobility, data on sitting-up was available only in 11 children in the CP group and 14 in the FTP group. Similarly, time to standing was calculated in 9 children of the CP group and 13 of the FTP group. The mean time to start walking was almost the same in both groups. It was recorded for 10 children in the CP group and 13 in the FTP group. The time at which the child starts to engage in playful activities was also lower in the FTP group; however, the difference was not significant. The time to return to school was affected by many social factors. Many children with perforation and perineal injury had left school since the time of first surgery and would only be able to join school once the next academic year begins, thus the average time to return to school could not be measured.

There were 8 complications in the CP group and 6 in the FTP group (Table 4). There was a statistically significant increase in wound infections in the CP group and paralytic ileus in the FTP group. Wound infections were managed with dressings and intravenous antibiotics. Paralytic ileus was managed by delayed feeding. One patient in the FTP group had anastomotic leak that required re-exploration and repeat colostomy formation. The leak was detected before the discharge of the child and thus led to an increase in the duration of primary hospital stay. The patient with fecal fistula in the CP group required readmission but could be managed conservatively. The patient with colonic dysmotility required bowel washes in the postoperative period.

## DISCUSSION

The primary duration of hospital stay and the overall duration of hospital stay (including readmissions) between the two groups were similar in the present study. These results cannot be compared with the results of the multiple studies done by Reismann et al[6,7] and Schukfeh et al[8], as they studied many different surgeries together and calculated the mean hospital stay due to all procedures. Thus they did not provide mean hospital stay for colostomy closure or bowel anastomosis. In the study by Sangkhathat et al[9] early enteral feeding after colostomy closure lead to significantly reduced post-operative stay, from an average of 6.1 days to 4.5 days. In our study, the mean postoperative stay for the FTP group was 4.33 days. Two out of the fifteen children in the FTP group were required to stay beyond post-operative day 5 (one was a case of an anastomotic leak who remained in hospital for 11 days, thus causing a skewed distribution and increase in the mean value). In the study by Mattioli et al[10], only 2 out of 46 required hospital stay beyond 5 days.

The mean pain scores were significantly lower in the FTP group (3.6, 2.93 and 2.93 for the day of surgery and postoperative days-1 and -2 respectively) compared to the CP group (5.33, 4.40 and 4.47 respectively). These results are better than those obtained by Reismann et al[6] where pain score during the immediate postoperative period was higher than 5 (4.2, 2.2, 2 for <4 years and 5.4, 4 and 2.3 for >4 years respectively). However, this study included a variety of surgeries and not just

Table 3. Return of Mobility

| Time to activity | CP Mean ± SD | FTP Mean ± SD | p-value |
|---|---|---|---|
| Sits up (hr) | 46.9 ± 24.9 | 14.6 ± 9.4 | <0.001* |
| Stands (hr) | 67.8 ± 29.3 | 23.2 ± 6.5 | <0.001* |
| Walks (d) | 3.4 ± 0.5 | 3.5 ± 5.0 | 0.493# |
| Plays (d) | 7.0 ± 5.4 | 5.3 ± 3.8 | 0.168# |

** Statistically significant, # Statistically not significant (using unpaired t-test), hr - hours, d - days. CP - Conventional protocol; FTP - Fast-track protocol*

Table 4: Incidence of various Complications

| Complications | CP n (%) | FTP n (%) | P Value |
|---|---|---|---|
| Ileus | 0 (0%) | 3 (20%) | 0.034* |
| Wound infection | 5 (33%) | 2 (13%) | 0.008* |
| Anastomotic leak | 0 (0%) | 1 (7%) | 0.155# |
| Vomiting | 1 (6.6%) | 0 (0%) | 0.155# |
| Faecal Fistula | 1 (6.6%) | 0 (0%) | 0.155# |
| Colonic dysmotility | 1 (6.6%) | 0 (0%) | 0.155# |

** Statistically significant, # Statistically not significant (using unpaired t-test), hr - hours, d - days. CP - Conventional protocol; FTP - Fast-track protocol*

colostomy closure. The children in the FTP group also required significantly lesser doses of rescue analgesia. This shows that preoperative counseling, epidural analgesia, the lack of drains and nasogastric tubes and active mobilization in the post-operative period lead to significant decrease in the pain scores. Only 1 child required removal of the epidural catheter on postoperative day-1 as it was blocked. Thus, insertion of epidural catheters is safe and effective even in young children and infants.

Return of bowel function was faster for the FTP group; however the difference was not statistically significant. In the FTP group, the mean time to tolerate liquid diet was 46.7 hrs and solid diet was 3 days. Flatus was passed in the meantime of 29 hrs and feces in 2.13 days. In the study by Mattioli et al[10] oral feeding and stool passage were achieved by post-operative day-1. In the study by Reismann et al[6], oral nutrition was achieved at a mean time of 15 hrs. However, this study included many surgeries which did not involve bowel handling, where it is possible to start feeds within a few hours of surgery. Sangkhathat et al[9] could achieve initiation of feeding within 19.7 hrs of surgery and full feeding by 45.5 hrs. Thus initiation of early feeding is feasible in the pediatric age group. Bowel function was also restored faster, probably owing to the avoidance of bowel preparation, initiation of early feeding, early mobilization and decreased pain in the FTP group.

Urinary retention is a known side-effect after epidural morphine administration. In our study, two patients of the FTP group and one of the CP group required catheterization due to urinary retention; however, this difference was not statistically significant. Thus, epidural morphine at a low dose of 30 μg/kg, does not cause significant urinary retention.

Two patients in the CP group required readmission, one for fecal fistula and one for wound infection (difference statistically not significant). This is in contrast to other studies of fast-track surgery in children[6,7,10] where readmissions were more amongst the fast-track patients.

The time taken to sit up and stand was significantly lower in the FTP group as compared to the CP group. The mean time to start walking was almost the same in both groups. The time at which the child starts to engage in playful activities was also lower in the FTP group; however, the difference was not significant. Reismann et al[6] could achieve mobilization in an average of 29.5 hrs, while in our study children were only walking after 3.46 days. This difference could be attributed to the fact that the former study included a variety of surgeries, some of which had faster convalescence than colostomy closure. It also could be attributed to cultural differences of the patient population. Many people still hold on to their traditional beliefs that strict bed rest is essential

for recovery after surgery. It was also observed that many children had not gone to school at all since the time of their previous illness (since the time of stoma formation). Thus they couldn't resume school immediately after surgery as many of them had lost one or two academic years and had to start school at the start of the forthcoming academic year.

The rate of complications was significantly higher for the CP group. There was a statistically significant increase in wound infections in the CP group and paralytic ileus in the FTP group. The increase in wound infections in the CP group may be related to the increase in stress response to surgery and the associated suppression of immunity. The increases incidence of ileus in the FTP group is probably related to the use of epidural morphine, which is known to decrease gastrointestinal motility.[3,5] One patient in the FTP group developed anastomotic leak that required re-exploration and repeat colostomy formation. The leak was detected before the discharge of the patient and thus led to an increase in the primary duration of hospital stay. In the CP group, vomiting, fecal fistula and colonic dysmotility were seen in one patient each. Reismann et al[7] defined complications associated with fast-track surgery as complications with a delay in diagnosis and treatment owing to early discharge. No such complications were seen in our study reported by Reismann et al.[6,7] In the study by Matioli et al [10] anastomotic leak and rectal pouch dehiscence was seen in one patient each. Thus, there were two complications among 46 patients. Our study thus replicates the complication rate of other studies.

Thus, in our study, the patients in the FTP group had a significant decrease in pain scores and need for rescue analgesia, a significantly shorter time to sit-up and stand and a significant decrease in overall complication rate. Patients in the FTP group had a significant decrease in wound infection and a significant increase in post-operative ileus (probably related to the use of epidural morphine). There was no significant differences in the duration of hospital stay, return of bowel function, need for catheterization, time taken to walk and play and readmission rate between the two groups. Thus, the entire gamut of traditional peri-operative care - including prolonged fasting, bowel preparation, use of drains and nasogastric tubes, intravenous analgesia and delayed feeding needs to be questioned. These measures are unnecessary even in the pediatric group of patients undergoing colonic surgeries. They increase the discomfort of the patient and do not lead to any significant improvement in results.

This study only included one type of surgery performed in the pediatric age group. More studies are required which evaluate the results in other abdominal surgeries too.

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Tropical surgery series

# Tropical Pyomyositis

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**Keywords**
Intramuscular Abscess
Myositis
Bacterial infection

**Abbreviations**
**PVL** - Panton-Valentine leukocidin
**SA** - Staphylococcus aureus
**TPM** - Tropical Pyomyositis

**Abstract**

*Tropical pyomyositis (TPM) is primarily an inflammatory disorder of skeletal muscles that is characterized by coagulative necrosis and superadded bacterial infection. It is common in tropical weather but not necessarily in a tropical country. Staphylococcus is the most common causative organism. TPM has 3 clinical stages. Local signs of inflammation such as redness and warmth are often deceptively missing. Streptococcal TPM is more aggressive disease. Mortality of stage-2 TPM is less than 2% while that of stage-3 approaches 20%. This article is a descriptive review of recent advances in this neglected tropical disease.*

## INTRODUCTION

**T**ropical pyomyositis (TPM) is primarily an inflammatory disorder of skeletal muscles that is characterized by coagulative necrosis and superadded bacterial infection. This definition excludes other suppurative infections of skeletal muscles that are secondary to penetrating injuries, osteomyelitis, septicemia or spread of infection from adjacent organs. It also excludes intermuscular abscesses, specific granulomatous infections such as tuberculosis and clostridial myolysis.[1] As its name implies, TPM is common in tropical climate; but not necessarily in tropical countries.

## HISTORY

In 1858, Gelle of France, described the first known case of TPM in his paper entitled "Suppurative myositis following muscle fatigue".[2] Subsequently De Salvia (1866) from Brazil[3] and Scriba (1885) from Tokyo[4] reported additional cases. In 1904, Miyake of Japan[5] established its pathogenesis by animal experiments. Interestingly, all these seminal papers of a disease named 'tropical' had come from temperate countries.

## NOMENCLATURE

TPM is known by a variety of synonyms. (Box 1) Native Africans called it Bungpagga disease or Bungura.[6] Rudolph Virchow is credited for coining the most popular term 'Tropical pyomyositis'.[1,7] In addition to these generic terms, site specific names such as ilio-psoas abscess, thigh or leg abscess and abscess of rectus abdominis are also frequently used. Some authors prefer 'Non-tropical Pyomyositis'[8] or 'Temperate Pyomyositis'[9] over TPM when the disease occurs in temperate countries. Such hypercritical semantic perfection is unnecessary for two reasons: (1) The adjective 'tropical' may mean 'tropical weather' rather than 'tropical geographic location'. High humidity combined with high environmental temperature appears to be essential for the

pathogenesis of TPM. For example, TPM is rare in Middle East countries with hot arid climate.[10] By corollary, TPM can occur in temperate countries when tropical weather prevails there. (2) Traditionally diseases are named on the basis of their high prevalence in a particular geographical locality. Rocky Mountain spotted fever, Kyasanur forest disease and Minamata disease are named with geographic identity, although they can occur at other places.

Box 1: Synonyms of Tropical Pyomyositis

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|---|
| Acute suppurative myositis |
| Bungpagga disease (Bungura) |
| Epidemic abscess |
| Myositis infectiosa |
| Myositis purulenta tropica |
| Myositis tropicans |
| Non-tropical pyomyositis |
| Primary intramuscular abscess |
| Primary pyomyositis |
| Primary suppurative (bacterial) myositis |
| Purulent infectious myositis |
| Pyomyositis tropicans |
| Spontaneous bacterial myositis |
| Suppurative myositis |
| Tropical myositis |
| Tropical skeletal muscle abscess |

## ETIO-PATHOGENESIS

Skeletal muscles are relatively immune to infections, because myoglobin in them avidly binds elemental iron that is essential for bacterial growth.[9] This is supported by the fact that intramuscular abscesses are extremely rare even in frank septicemia.[11,12] In a series of 201 cases of staphylococcal bacteremia, none developed TPM.[11] Among 327 cases of fatal staphylococcal sepsis, [12] there were only 2 cases of TPM, even which appears to be the cause rather than the effect of septicemia. Intrigued by these facts, Miyake[5] conducted rabbit experiments by intravenous injection of sublethal doses of *Staphylococcus aureus (SA)*. Intramuscular abscess did not occur unless the muscle was made vulnerable by prior trauma such as pinching or electrical stimulation. It was hypothesized that muscle injury liberates sequestrated iron from myoglobin. Iron needed for microbial growth is also likely to be released from hemoglobin of traumatic intramuscular hematomas. Miyake's experiment proved that two concomitant factors are essential to cause TPM: (1). Presence of bacteremia, (2) A pre-existing muscle injury or defect that localizes the circulating bacteria. Miyake's conclusion is consistent with Knudson's two-hit hypothesis of pathogenesis which says that two concomitant factors are essential to cause any disease. Several such predisposing factors have been identified. (Box 2)

In tropical climates, high salt content of sweat and its inhibitory effect promotes selective colonization of skin with SA. This may explain as to why TPM is common in hot humid periods. Predisposing muscle damage may be traumatic or non-traumatic. Severity of injury may vary from unrecognizable subtle tear of fibers to overt penetrating injuries. Subclinical injuries as a result of strenuous exercise or sports activities occurring in 5-39% of TPM may actually be ignored as 'muscular fatigue'.[13, 14, 15]

Skeletal muscle damage may also occur in dietary deficiencies and infectious diseases. In Japan, the incidence of TPM dropped with the disappearance of dry-beriberi which is known to cause hyaline degeneration of muscles. TPM is also common in areas where polished rice devoid of thiamine is eaten. By animal experiments, Osawa confirmed the role of vitamin $B_1$ deficiency in the etiology of TPM.[16,17]. In Uganda, protein energy malnutrition and muscle wasting was noted in 94% of TPM.[18] Tiny intramuscular hematomas of vitamin-C deficiency (Scurvy) was suspected to favor bacterial invasion; but it could not be proved.[17,19] Although deficiencies of Vitamin E, Vitamin A and selenium were shown to cause TPM in cattle, none is proved in human beings.[14, 17, 19] In Japan, TPM is common during the seasons of sweet potato cultivation. In India, it is common during the

Box 2: Predisposing factors of Tropical Pyomyositis

**Source infection of bacteremia**

- Tropical weather favoring selective colonization of the skin with *SA*
- Pyoderma (e.g. boils, eczema, chicken pox, paronychia)
- Otitis media
- Pneumonia
- Traumatic wounds
- Crohn's disease, Ulcerative colitis

***I*mmuno-compromised state that promotes bacteremia**

- Cancers (e.g. Leukemia, lymphoma) and their chemotherapy
- Type-1 Diabetes mellitus
- HIV infection
- Immunosuppressive / myelosuppressive medications (e.g. Corticosteroids, post-transplant medications, anticancer drugs)
- Chronic liver or renal failure
- Malnutrition
- Intravenous drug abuse
- Agammaglobulinemia
- Congenital immune deficiency disorders (e.g. $C_3$ complement deficiency, Myeloperoxidase deficiency, IgM deficiency)
- Myelodysplasia
- Atopic dermatitis

**Muscle defects that facilitate localization of circulating bacteria**

- Hemoglobinopathies (Beta-Thalassemia, Sickle cell anemia)
- Dermatomyositis
- Polyangitis
- Connective tissue disorders (e.g. Rheumatoid disease, systemic sclerosis, Systemic lupus erythematosis, Felty's disease)
- Syphilis
- HIV myositis
- Viral myositis of exanthematous fevers (e.g. Influenza, coxsackie virus B, measles, herpes, arenavirus, picornavirus, arbovirus, leptospirosis )
- Blunt trauma (e.g. bicycle injuries and sports injuries)
- Penetrating injuries (e.g. Intramuscular injections and surgical incisions)
- Strenuous exercise, sports injuries, muscle fatigue
- Intramuscular parasites (e.g. *Trichuris*, *Cysticercus*, *Ancylostoma*, *Dracunculus*, Filarial worm, *Trypanosoma*, *Toxocara*)

*SA - Staphylococcus aureus; HIV - Human Immunodefeciency Virus; IgM - Immunoglobilin-M*

harvesting season of rice and wheat. In New Guinea and Uganda, TPM is common after pork feast. From these observations, Shepherd[14] hypothesized that abrupt change in dietary habit may precipitate bacteremia by altering the colonic flora. High quality evidences are missing to support this hypothesis.

Several infectious diseases are known to cause pre-disposing muscle damage of TPM. Coinciding

of mosquito breeding season, malarial epidemic and TPM outbreak raised a suspicion that TPM could be a vector born muscle pathology.[14,17] Zenker's degeneration myopathy of typhoid and myopathy of human immuno-deficiency virus (HIV) are incriminated in the pathogenesis of TPM.[14,20] The association between HIV and TPM appears to be real with an odds ratio of 4.82.[20] Eosinophilia, which is unusual in acute bacterial infection, is seen in 5% of TPM. This prompted a search for parasitic etiology. In 7-63% of TPM patients stool was positive for *Ancylostoma* eggs.[21] Intra-muscular helminthes such as the hookworms, *Trichuris*, cysticercus and *Dracunculus* were thought to render the muscle susceptible for bacterial invasion.[17,21,22] However, microscopic examination of affected muscle or pus did not show any evidence of worm infestations. Often, a predisposing factor may not be easily evident.[17]

Exact mechanism, as to how the circulating bacteria get localized at the site of muscle damage, is not known. Trogocytosis is recently proposed to play a role. It is a phenomenon by which circulating lymphocytes bind bacteria by immune complex conjugation and transmit them to the recipient myocytes.[15,23] Increased risk of TPM in patients receiving monoclonal antibodies such as certolizumab, tocilizumab and infliximab may be attributed to immuno-complex conjugation analogous to trogocytosis.[26]

Clinical severity of TPM is linked to Panton-Valen tine leukocidin (PVL), an exotoxin secreted by certain strains of *Staphylococcus* for their survival advantage.[24] It causes leukocyte-mediated tissue destruction and necrosis.[25] PVL, by binding CD45 and C5a receptor, causes lysis of granulocytes and release of proteolytic enzymes which leads to autodigestion of tissues.[26] PVL is now recognized as a critically essential factor in the pathogenesis and outcome of TPM in tropical countries.[24] PVL positive SA cause severe disease and leads to prolonged hospitalization.[27] PVL positive SA can cause TPM even without antecedent muscle damage.[27]

## DEMOGRAPHY

True TPM is relatively rare even in tropical countries.[14] It has been reported from almost all countries, with increased frequency from tropical areas.[15] A history of recent travel to a tropical country is obtained only in 9% of cases.[9,17] Its incidence varies from 1-in-1000 (tropics) to 1-in-5000 (non-tropics) population per year.[21] TPM constitutes 1-4% of all admissions in African hospitals.[21] In Ugandan hospitals as many as 1000 cases per year were seen.[14] Within North America it is more common in tropical belt (e.g. Mexico and Texas) than in Canada or Iowa.[25] There are some data to suggest that the incidence is falling in tropics while increasing in temperate countries. For example, annual incidence of TPM in Mulago Hospital, Kampala fell from 250 to 71 during 1948 to 1961.[28] On the contrary, its incidence in an Australian hospital increased from 2.04 cases per 10,000 emergency admission in 2002, to 8.7 cases in 2012 (with a peak of 13.5 cases in 2008).[29] Increased awareness about TPM, better diagnostic imaging, effect of global warming, ease of international travels and antibiotic misuse could be the reason for this changing incidence.

Slight male preponderance of TPM in tropics (M:F - 1.4:1 to 2.3:1) is probably attributable to gender related bulkiness of skeletal muscles or increased chances of sports injuries in boys.[25,30] In non-tropical areas, male-female ratio is as high as 4:1.[31] Cook noted a chronological trend of narrowing sex-ratio. He found male-female ratio of 5.5:1 in 1948 which became 2.7:1 in 1961.[28]

The two peaks of age incidence are 2-9 years and 20-40 years.[10,14] In pediatric age group, Royston noted 51% occurring between 3-6 years of age while 31% occur between 9-12 years.[32] No age

including neonatal period is immune to TPM.[33,34] The youngest patient reported in literature was a 6-days-old newborn.[33] Fewer than 7 cases have been reported in neonates.[33,35]

Different races appear to vary in their susceptibility to TPM, although no genetic correlation could be proved. Seventh Day Adventist of Solomon Islands are said to be immune to TPM as they do not eat pork.[14,36] In tropics, Caucasians migrants are more affected than natives.[14,17] Even in the same race different patterns of dietary habit is said to be associated with varying frequency of TPM. For example, among Africans incidence of TPM is high in Eastern Uganda where the staple diet is cassava and sweet potato while it is low in Western Kenya where maize is the main course.[14]

TPM is common during July to October when humidity is high.[30] It coincides with harvesting season of rice and guinea corn.[25,37] It is rarely seen above an altitude of 4000 feet from sea level.[37]

## PATHOLOGY

Involvement of single muscle is more common than multifocal lesions (10-40%).[21,22,25] Right handedness of majority population and hence the increased risk of subclinical injury to muscles could be the reason behind higher incidence of TPM on the right side (R:L - 3:2).[1,15,17] Frequency of individual muscles affected is directly proportional to the bulkiness of the muscle and to strenuous activities.[21,25,30,38](Fig 1) However, even small muscles are not spared. Shepherd cited a case of TPM of orbital muscles that presented as proptosis.[14] The amount of pus drained from a single lesion may be as high as 300 ml to 2 liters.[21,26] Despite bacterial nature of the infection, regional lymph nodes are seldom enlarged.[14]

Histologically the affected muscles show Zenker's degeneration with infiltration of acute inflammatory cells including monocytes and eosinophils. Electron microscopy of the affected muscles show degenerative changes in apparently healthy looking areas well away from the lesion.[39] These

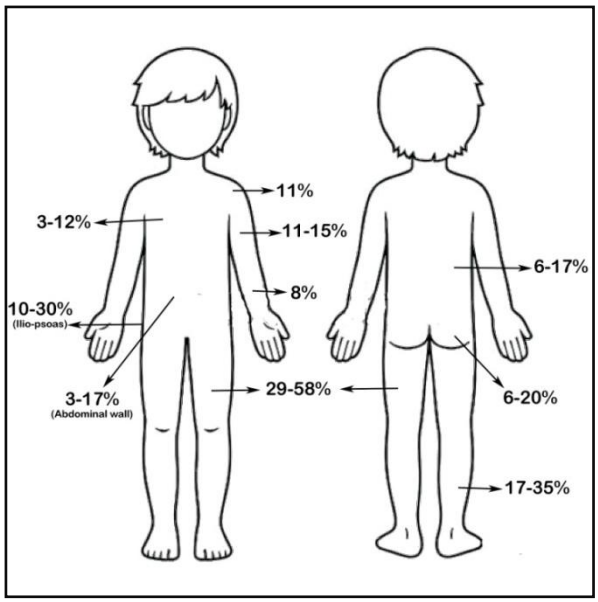


**Fig 1:** *Anatomical distribution of tropical pyomyositis in children (Data from Chinda[38] Ansaloni[21] Verma[25,30])*

changes are not easily appreciated in light microscopy.[39] Even distant muscles such as the myocardium may show submicroscopic changes.[39] This indicates that TPM is primarily a degenerative disease of muscle to which hematogenous bacterial infection is superadded.

## MICROBIOLOGY

*Staphylococcus aureus* (*SA*) is the major offender in 70-90% cases.[40] Phage type-2 SA, recovered from 60% of TPM, was originally thought to be of pathogenic significance;[40] but subsequent studies did not support this hypothesis. Other microbes occasionally reported in TPM include *Streptococcus* (5-10%),[36] *Pneumococcus,*[41] *Enterococcus,*[42] *Klebsilella,*[43] *Proteus, Escherichia,*[44] *Salmonella,*[45] *Citrobacter, Enterobacter, Morganella, Haemophilus, Aeromonas, Serratia, Pseudo-*

*monas, Nocardia* and *Yersinia*.[15] Often mixed infection of anaerobes such as *Clostridium difficile*, *Peptostreptococci* , *Bacteroides fragilis, Fusobacterium*, *Prevotella* and *Veillonella* is seen.[15,46] Although other microbes such as *Mycobacterium tuberculosis*, *Mycobacterium avis*, *Candida albicans* and *Cryptococcus neoformans* have been reported in the literature,[15] by strict definition those specific infections should not be included under TPM.

In recent years, an increasing proportion (46-63%) of *Staphylococcal* isolates are community acquired; of which 75% are methicillin resistant SA (MRSA) and 25% are methicillin susceptible SA (MSSA).[27,30] PVL positive strains and USA300 clones affect relatively younger patients, have shorter duration of bacteremia and cause larger abscess than PVL negative SA.[27]

## CLASSIFICATION

TPM is classified in a number of ways such as 'primary *versus* secondary'; 'axial (paraspinal or thoraco-abdominal) *versus* appendageal (limbs)'; and 'focal *versus* multiple'. These classifications are not popular as they lack clinical significance. However, tropical TPM significantly differ from non-tropical TPM.[9] In temperate countries TPM occurs mostly in immuno-compromised adults (60%) while in tropics it predominantly affects healthy young children. Male preponderance in temperate climate (4:1) is much exaggerated than that of tropics (1.4:1). Rarity of TPM in temperate countries often leads to diagnostic delays.[47] The usual delay in diagnosis is 12-16 days while it may be as long as 3 months.[21] Christin reported North American patients with an unusual delay of 1 year.[17] Blood cultures are more often positive (35%) in temperate countries than in tropical centers (5%).[1,9] *Staphylococcus* is more common in tropics (90%) while it accounts for only 75% in temperate countries.[9] PVL strains are more often seen in tropical TPM than in non-tropical TPM.[48]

## CLINICAL FEATURES AND STAGING

Three distinct clinical stages of TPM have been described.[1,17,21,25,31] They are as follows:

### *Stage 1 (Stage of invasive myositis)*

In this stage bacterial invasion of muscle causes features of inflammation which include low grade fever, local muscle pain, fusiform diffuse swelling of the involved muscle, muscle spasm and flexion deformity of the associated joint. This stage usually lasts for 7-21 days. Symptoms are often less dramatic and vague (e.g. dull muscle ache, anorexia, general malaise and cramps). The affected muscle may be woody hard in consistency due to tense edema within investing fascia (epimysium). Tenderness of the involved muscle may be mild or absent. As the inflammation is deep to deep-fascia, skin over the lesion may look deceptively normal.(Fig 2) Diagnostic needle aspiration would be negative. Very few patients (<5%) present at this stage and they are often misdiagnosed due to their non-specific clinical features.[31]

**Fig 2.** *Clinical appearance of tropical pyomyositis of the right thigh. Overlying skin may deceptively look normal. (Representative illustration generated by AI technology using 'wepik' software)*

### *Stage 2 (Stage of suppuration / abscess)*

In this stage the localized infection progresses to suppuration of muscle resulting in an abscess. Inflammatory signs will be more prominent (e.g. high grade fevers with chills, local tenderness, severe crippling of muscle function and sick

appearance). Pseudofluctuation of overlying muscle belly should not be mistaken for pus collection. It lasts for 1-12 days. Diagnostic aspiration with a wide bore needle would yield pus. Overlying skin may still look deceptively normal. Rarely, the skin may be warm, red, edematous and shiny. About 90% of patients present at this stage.[31]

### *Stage 3 (Stage of dissemination)*

Clinical features of this stage are similar to stage-2 with the addition of systemic manifestations of sepsis and circulatory shock. The muscle infection may spread to adjacent and distant organs. Mortality is usually 2-10%. About 2-3% of children present at this stage. [31]

## DIFFERENTIAL DIAGNOSIS

Vague symptoms of stage-1 are easily mistaken for a variety of conditions including non-specific myalgia, sciatica, backache, viral fever, poliomyelitis, dermatomyositis, toxoplasmosis, deep vein thrombosis, thrombophlebitis, trypanosomiasis and Perthes disease.[15,21,25] Morison's aphorism says, "Cellulitis occurring in children is never primarily in the cellular tissues but secondary to an underlying bone infection".[49] Hence, many a TPM are mistaken for osteomyelitis or vice versa. Nearly 32% of children suspected to have septic arthritis of hip turned out to be TPM. In true arthritis, joint movements are restricted in all directions while in TPM it is restricted in only one axis.

Swelling of stage-2 lesion may be mistaken for muscle hematoma, soft tissue sarcoma, nodular fasciitis or infantile fibromatosis.[50] Conversely rhabdomyosarcoma has been mistaken for TPM.[51] Psoas abscess of right side may mimic acute appendicitis or appendicular abscess.[52]

## INVESTIGATIONS

Blood investigations[21,25,14] may show leucocytosis (49%), eosinophilia (5%), elevated ESR (91%) and increased C-reactive protein level (93%). Blood cultures are rarely (5%) positive. They are more likely to be positive if the organism is *Streptococcus* rather than *Staphylococcus*.[17] Pus cultures are positive in 10-40% cases.[25,30] High rate of negative culture is attributed to partial treatment with antibiotics prior to sampling. Recently, polymerase chain reaction technique has been introduced to overcome this problem and to increase the diagnostic yield.[15, 53] Coexisting HIV in 31% and intestinal hookworm in 17-63% should also be investigated.[20,21]

Inflammatory obliterated muscle planes in plain radiographs are highly non-specific.[54] However, x-rays may be useful in excluding osteomyelitis as a differential diagnosis. Ultrasonography (USG) is useful in differentiating stage-1 and stage-2 of TPM.[32,55] Hypo-echoic shadows within the muscle and floating debris or gas bubbles are diagnostic of abscess. However, it must be remembered that sometimes edematous muscle of stage-1 TPM may sonographically mimic an abscess. USG is often useful in guiding needle aspiration and in excluding septic arthritis.

Computed Tomography may show low attenuation of abscess collection, gas bubbles within the muscle and peripheral contrast enhancement of abscess cavity. Point-of-care ultrasonography (POCUS) is comparatively better than CT in diagnosing TPM.[56] Magnetic resonance imaging (MRI) is more sensitive than USG and CT scan.[57,58,59,60] It is claimed to effective in diagnosing TPM within 3-5 days of onset.[57] Hyperintense signals in $T_2$ weighted images and hyperintense rim on enhanced $T_1$ weighted images are characteristic of intramuscular abscess. Often the lesion is larger in MRI than that is appreciated clinically. Gallium scintigraphy[61,62], although a sensitive method, lack precision and specificity. Positron emission tomography (PET-CT) or Single photon emission computed tomography (SPECT) scans are said to be useful in multiple or occult

lesions.[63,64] In resource poor settings, the diagnosis of abscess is often confirmed by repeated diagnostic aspiration with a wide bore (16G or larger) needle rather than by imaging.

Despite extensive myolysis, muscle enzymes such as lactate dehydrogenase (LDH), creatine phosphokinase (CPK), and aldolase are not elevated. If they are elevated, it may suggest either multifocal TPM[65] or an alternate diagnosis such as polymyositis. In school going children TPM occurring as a complication of influenza is associated with marked rise in CPK.[30] Electromyography (EMG) may show short duration, low amplitude polyphasic potentials; but it is not recommended in fear of spreading the infection.

## TREATMENT

Stage-1 TPM is treated with appropriate antibiotics that can cover *Staphylococcus*. Initially antibiotics are given intravenously for 7-10 days followed by oral antibiotics. Antibiotics are usually needed for prolonged period (average 3 to 4 weeks with a maximum of 94 days reported by Christin).[15,17,35] Combination therapy is preferable over monotherapy to avoid development of bacterial resistance. Cloxacillin, nafcillin, oxacillin, flucloxacillin, ceftazidime, cefepime, piperacillin-tazobactam, carbapenem, aminoglycosides, third generation cephalosporins, clindamycin, penicillin are the commonly used drugs. Anaerobic coverage by metronidazole is given during the first few days of therapy. Vancomycin, linezolid, teicoplanin and daptomycin are reserved for Methicillin or Vancomycin resistant *SA* (MRSA or VRSA respectively). Quinolones (ciprofloxacin, ofloxacin), doxycycline and fusidic acid are rescue drugs that are rarely used in older children. In case of multidrug resistant organism, rifampicin may be considered. There are some evidences to suggest that macrolides, lincosamides, rifampicin or oxazolidinone inhibit production of PVL, thereby facilitate quicker recovery.[66] In stage-3 TPM intravenous immunoglobulins may reduce the mortality of toxic shock syndrome.

When pus is localized, surgical drainage is essential in addition to antibiotics. Although occasional papers[67] reported success with antibiotics alone without surgical drainage of pus, author of this review does not subscribe to that view. The standard surgical dictum *"Ubi pus, ibi evacua"* should never be violated. Usually open drainage is preferred over percutaneous tube drainage or image-guided therapeutic aspiration. Multiple loculi of the abscess cavity cannot be effectively broken by percutaneous drainage techniques. A drainage tube is left *in situ* as long as it drains. Surgical access is determined by the location and size of abscess. Ilio-psoas abscesses are drained either by transabdominal extraperitoneal route or by laparoscopy.[68]

Mutilating surgeries[69,70] such as the excision of rectus femoris and amputation are no longer required. Such radical operations may be occasionally needed especially in severe streptococcal TPM.[70]

Failure of clinical symptoms to resolve with antibiotic treatment may be due to incomplete drainage of pus, occult abscess elsewhere, antibiotic resistance or drug-induced fever. Endpoints of antibiotic therapy include resolution of clinical symptoms, fall of erythrocyte sedimentation rate (ESR) to normal level, return of C-reactive protein to normalcy.

Topical nasal application of mupirocin cream is sometimes used as a therapeutic adjunct or prophylaxis to eradicate nasal carrier state of *Staphylococcus*.[1]

## COMPLICATIONS AND OUTCOME

Although a majority of patients recover uneventfully without any significant residual deformity or recurrence, mortality of stage-2 TPM is 0.8-2%

and that of stage-3 is 2-23%.[10, 20] Streptococcal TPM is more serious than staphylococcal TPM.[34] PVL positive SA causes more deaths than PVL negative infections.[71] Average hospital stay is 14 days (range 7 days to 90 days).[26] Recurrences are exceptionally rare. Complications of TPM vary between 9 to 66%.[15] They include osteomyelitis (5-41%), septic or reactive arthritis (7-25%), pneumonia (18%), septicemia (3-5%), toxic shock syndrome (2%), multi-organ dysfunction syndrome (<1%), spinal epidural abscess, pyopericardium, myocarditis or endocarditis, meningitis, brain abscess and empyema thorax.[25] Osteomyelitis may be contiguous (adjacent bone) or hematogenous (distant bone). Compartment syndrome and compression neuropathy [72] have also been reported. Myoglobinuria due to extensive myolysis may precipitate renal failure. [14]

(Endnote: Peculiarities of site specific pyomyositis will be addressed separately in this series. Randomized controlled trials are non-existent and the only one meta-analysis available examines the association of HIV and TPM. Thus, recommendations made in this article are derived from case series and isolated case reports)

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Clinical Research

# Study of Short-Term Outcomes of Targeted Therapy in Operated Patients of Hirschsprung Disease Who Were Found To Have Poor Functional Outcome at Initial Assessment

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**Keywords**
Hirschsprung Disease
Bowel Function Scoring
Fecal Incontinence
Constipation

**Abbreviations**
**BFS** - Bowel function score
**DPT** - Duhamel pull-through
**EUA** - Examination under anesthesia
**HD** - Hirschsprung Disease
**HIC** - High income countries
**LMIC** - Low- middle- income countries
**QoL** - Quality of life
**TCA** - Total colon aganglionosis

***Abstract***

***Background:*** *Long-term morbidity and functional bowel problems after surgical treatment of HD are significant. There is no consensus regarding the most appropriate scoring system for the assessment of functional outcomes. Also, there is a paucity of data from low- and middle-income countries (LMIC).*

***Materials and Methods:*** *This prospective interventional study included 31 children, aged 4 to 18 years, who had undergone definitive surgery for Hirschsprung disease (HD) between 2011 and 2020. Among the cohort, those with low bowel function score BFS (<17) were selected for targeted therapy of constipation, soiling, or both.*

***Results:*** *There were 19 patients with BFS < 17. Male-female ratio was 17:2. The mean age at enrollment was 6 yrs (range 4.3 - 12 yrs). Fifteen (79%) of then had classical HD, while 4 (21%) had long-segment HD. The mean time elapsed since surgery was 3.86 ± 2.37 years. Twelve (63%) of them had constipation, while 7 (37%) had both constipation and soiling and none had exclusive soiling. The mean BFS before and after targeted therapy was 15 ± 1.05 (range 13-16) and 18.21 ± 0.63 (range 17-19) respectively. After intervention, in all of them BFS improved ≥17.*

***Conclusions:*** *HD has significant problems for many years after definite surgery. Using BFS, we can identify patients who need early targeted interventions including medical and surgical therapy. The extent of aganglionic bowel, age at the initial presentation and gender do not appear to affect the final outcome. Short-term 3-month targeted therapy was beneficial in those with BFS<17.*

## INTRODUCTION

Hirschsprung disease (HD), congenital absence of ganglion cells of the distal bowel, is one of the most common surgical conditions in the pediatric age group.[1,2] Surgical resection of poorly functioning aganglionic segment and an anastomosis of a normally innervated proximal bowel to the distal rectum is the definitive treatment of HD. Final outcome and long-term function do not differ significantly between various surgical operations such as the Swenson's procto-colectomy, Duhamel pull-through (DPT), Soave's operation and laparoscope-assisted endo-rectal pull-through.[3]

Jarvi et al. found that long-term morbidity and functional bowel problems are significant after definitive treatment of HD. Fecal incontinence and constipation are troublesome postoperative complications. Obstructive symptoms may be due to residual aganglionic segment, stricture, or dysmotility, whereas fecal incontinence may be due to operative injury to anal sphincter, inappropriately low colo-anal anastomosis or overflow soiling.[4]

Various factors that may affect the quality of life (QoL) including the age at surgery, sex, type of surgery operation, level of aganglionosis, initial stoma diversion, enterocolitis episodes, and bowel dysfunction have been studied. None of them other than bowel dysfunction was found to impact the QOL.[5]

Although HD is a disease of childhood, its final outcome continues to evolve well into adolescence and adulthood. A combination of several parameters must be carefully assessed at specified intervals to identify patients who are liable to have mid-and long- term complications and those who will benefit significantly from early intervention.

To date, there is no consensus regarding the most appropriate scoring system for assessing the functional outcomes in these patients. Some authors like Hartman[6] use generic questionnaires while others like Meinds use Rome- IV criteria of constipation.[7] Some authors like Moore et al[8] from South Africa and Gabriella et al[9] from Indonesia have used anthropometric data as an index to compare the long-term effect of HD surgeries on the child's growth and development by comparing them with standardized growth charts. However mere physical growth is an insufficient indicator of overall development.

Another important point is that most of these studies were done in high-income countries (HIC) where the standards of ancillary services are excellent. A study by Kumar et al from Vellore, India examined the functional outcomes and QOL in HD; but did not find significant differences between the patient cohort and healthy controls. This was a cross-sectional study and it did not evaluate the long-term result of targeted interventions.[10]

Thus, a critical assessment of mid- and long-term complications in patients operated for HD in LMIC is lacking. We hypothesized that patients with low bowel function score (BFS) during initial assessment, if provided with targeted intervention without delay, can lead near-normal lives well into adulthood. This study was conducted to examine this hypothesis.

## MATERIAL AND METHODS

A prospective interventional study of children treated for HD was performed after receiving clearance from the institutional ethical committee. The patients aged 4 to 18 years who had undergone definitive surgery for HD at the Pediatric Surgery Department of our tertiary center from 2011 to 2020 were recruited for the study after due approval of Institutional Ethics Committee. Those who underwent definitive surgery within 6 months of study enrollment and those who were not willing were excluded from the study. Some of them had temporizing diverting

colostomy done elsewhere. Patients having total colonic aganglionosis (TCA), ultra-short HD (anal achalasia), severe neurological deficit, permanent stoma or hypothyroidism were excluded from the study. Of the patients were re-operated for HD over seven years (July 2016- November 2022), 56 could be contacted and only 31 patients agreed to be enrolled in the study. Previous records of operated patients were retrieved and reviewed. Patients were convened to the hospital for physical examination. Those who could not visit the hospital were contacted by phone and the conversation was recorded. Those who experienced problems related to surgery were encouraged to visit the hospital. Written informed consent was obtained from each participant. Details regarding the length of aganglionic bowel, nature of surgical treatment, episodes of enterocolitis, complications, and any additional surgical interventions needed were noted from previous records.

HD was classified as: (a) classical - if the aganglionic region was limited to the rectum and sigmoid colon; (b) long segment - if it extended proximal to the sigmoid colon but not the entire colon. The definition of constipation used was according to the criteria proposed by Drossman et al.[19] Constipation was defined as straining at the stools more than 25% of the time or less than 3 stools per week. Fecal soiling was defined as small amounts of feces having to be scrubbed off the underclothing at least once a month.[20] Incontinence was defined as the inability to control bowel movements causing the child of age $\geq$4yrs to repeatedly pass stools in inappropriate places.[21] Enterocolitis was diagnosed by the combination of abdominal distension, diarrhea or bloody stools, vomiting, and fever. Mid-term complications were defined as those occurring between 6 months - 2 years following definitive surgery. Long-term complications included those occurring after 2 years of surgery.

All patients were assessed using Bowel Function Score (BFS) questionnaire (Appendix-1). BFS is a 20-point score which includes various parameters like constipation, soiling, urgency, frequency of defecation, fecal accidents, and social problems. All those with BFS $<$ 17 (n=19) were categorized as poor outcome patients and labeled as 'index cases. Among these, 16 (84%) developed symptoms after 6 months of surgery and 3 (15%) after 2 years of surgery.

All the index cases underwent barium enema imaging of colon. (Fig 1) Those presented with exclusive constipation had examination under anesthesia (EUA), while those with both constipation and soiling had both EUA as well as anorectal manometry. All were offered with targeted therapy for the condition diagnosed on investigations. (Fig 2 and 3) The effect of the targeted therapy was studied after 3 months and the improvement in BFS, if any, was recorded.

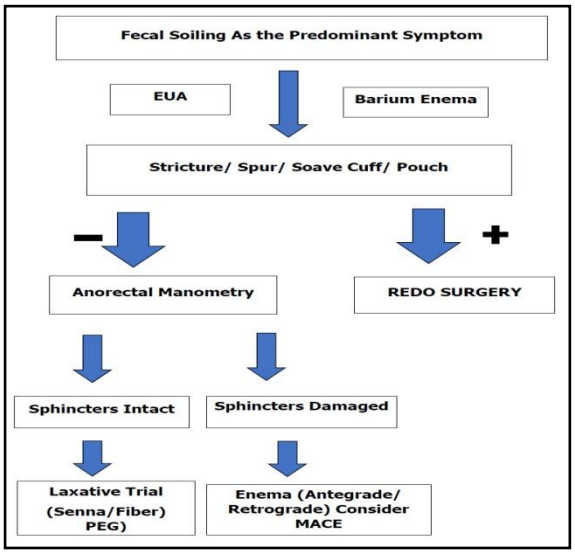


**Fig 1.** *Scheme of evaluation*

## RESULTS

Of the 31 enrolled patients, 90% (n=28) were males and 10% (n=3) were females. The average age at enrolment was 7.4 years in males and 6.3 years in females. The mean age at initial contact for definitive treatment was 3.6 $\pm$ 3.5 yrs (range: 4 mo to 11 yrs). Of them, 23 (74%) had classical HD, while 8 patients (26%) had long-segment HD.

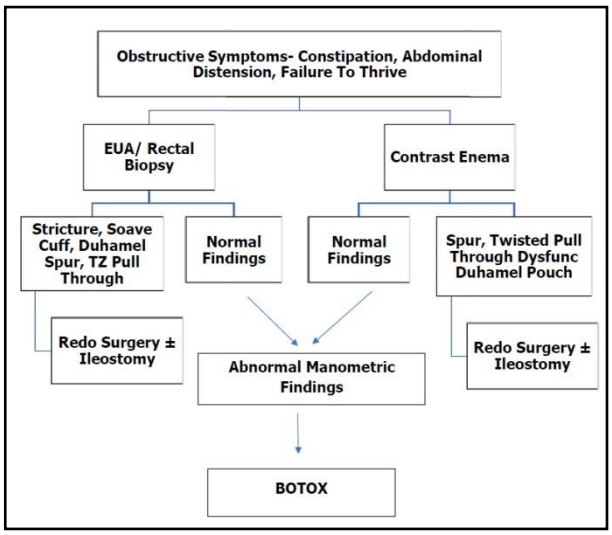


**Fig 2.** *Algorithm of targeted therapy for Bowel Function Score <17 with constipation*

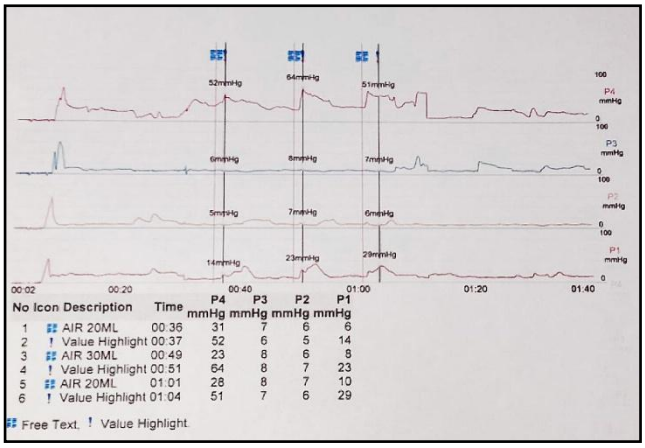


**Fig 3.** *Anorectal manometry of 12 year male patient (Pre-treatment BFS - 16)*

The mean time elapsed since definitive surgery was 5.8 ± 2.4 yrs.

The mean BFS of 31 enrolled patients at presentation was 18.8 ± 0.8. Among them 19 had BFS <17. There was no difference in BFS of males and females (14.94 ±1.09 vs. 15.50 ±0.71); however meaningful analysis was not possible as there were only 3 girls.

The mean BFS of the 19 'index cases' before giving targeted therapy was 15 ± 1.05 (range 13-16). The mean age of symptomatic presentation of the index cases was 1.42 yrs. Fifteen (79%) of the index cases had classical HD, while 4 (21%) had long-segment HD. BFS of classical versus long-segment disease (15.07±0.96 vs. 14.75±1.50 respectively) did not differ significantly. All 19 patients had undergone DPT as the definitive surgery. The mean time elapsed since surgery was 3.86±2.37 years. Twelve (63%) of the index cases had exclusive constipation, 7 (37%) had both constipation and soiling and none had exclusive soiling.

Barium contrast enema in all 19 index cases was normal. (Fig.4) Seventeen (89.5%) of the index cases had normal EUA findings, while 2 (10.5%) were found to have spur-on EUA; these 2 patients underwent 'spur division' at the same sitting. The other 17 patients (89.5%) were managed medically with dietary modifications (fiber rich diet such as salad, green vegetables), adequate fluid intake, laxatives, and water-soluble fibers to increase the bulk of the stool. The mean BFS after targeted therapy was 18.21 ± 0.63 (range 17-19). All the index cases improved after targeted therapy with a BFS ≥17.

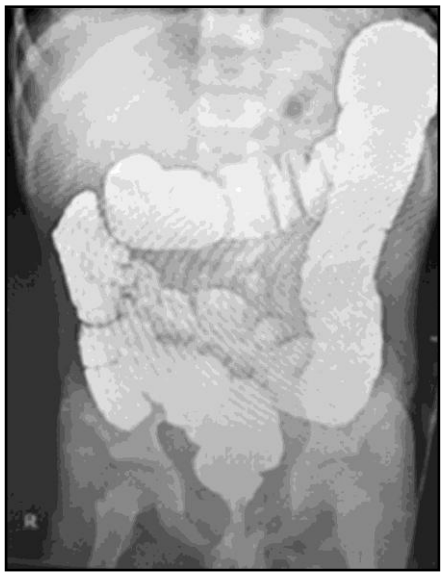

**Fig 4.** *Barium enema in 5-yr-old boy (Pre-treatment BFS 16)*

## DISCUSSION

Duhamel introduced a new operative technique for Hirschsprung's disease in infants in 1956, his

procedure has gained worldwide acceptance. Heij et al. performed a systematic follow-up after DPT and confirmed a considerably high incidence of mid- and long-term complications. They observed that 17 out of 63 suffered some degree of incontinence, and 22 experienced soiling and/or constipation.[11]

Constipation is a major problem after DPT due to a large capacity reservoir partially consisting of the aganglionic bowel. Rescorla et al showed that 27% of DPT patients used enemas or stool softeners, and 8% had severe constipation.[12] In the current study also 61% of patients had significant problems after DPT.

Among the 19 index patients with low BFS, the mean pre-therapy BFS was 15.05 (range 13-16). This implies that patients who had undergone DPT have significant problem several months after the operation.

All index cases were evaluated for improvement, after 3 months of targeted therapy. It was heartening to note that the targeted therapy was effective in managing mid- and long-term complications in all of them.

Considerable literature is available for short- and mid-term complications of HD, but there is very little literature regarding long-term complications especially from LMIC. Ying et al found the prevalence of fecal incontinence, constipation, and bladder dysfunction after 10 yrs of definitive surgery was 20%, 14%, and 7% respectively.[13] Overall pooled BFS and QoL scores showed satisfactory results.

When analyzing the outcomes definitive treatment of HD, various surgical techniques must be taken into consideration. Various studies have compared outcomes with respect to DPT, Swenson, Soave, Laparoscopic endorectal pull through (LERPT), and transanal endorectal pull through (TERPT). However, there is no clear advantage of one procedure over the other. Different risks and benefits are associated with these surgeries.

Since most of the surgeries performed at our center were DPT, we had no opportunity to study the outcomes of other surgical techniques. As regard to DPT in this study, 12 patients had constipation while 9 had constipation with soiling, and none had soiling alone. The symptom of only soiling (called true incontinence) signifies sphincter damage during surgery. Probably no other complication has a greater impact on the QoL than the incontinence. Unfortunately, incontinence rates are often not reported in many of the larger series. Others failed to make a distinction between occasional soiling and significant incontinence.

It is difficult to attribute a cause of high incontinence rates in some series. Comparison of series is difficult as the technique, expertise and meticulousness of stool history vary considerably between them.[14] Since none of our patients had 'true incontinence' it can be said the DPT is a relatively a safe procedure to be done by experienced surgeons.

In our study, the final functional outcome after DPT for HD was independent of the length of aganglionosis. Contrary to this, Shu et al[15] found that among the 12 out of 58 (21%) patients who were symptomatic, one-third had aganglionic segment less than 30 cm and two-third had aganglionic bowel more than 30 cm, whereas in the 46 asymptomatic patients, 91% (n=42) had less than 30 cm aganglionic segment, and 4 had 30 cm or longer aganglionic segment. This difference was statistically significant thereby suggesting that the extent of HD has an impact on the outcome. Similarly, Moore et al[16] also found a significant difference in outcomes of patients with classical HD versus long-segment HD versus TCA. Of the 178 responders, 123 (69%) had a classical HD, 41 (23%) had a long segment HD, and 14 (8%) had TCA. In 29 patients with postoperative

complications, 14 had long segments HD (48.2%), and 5 (17.2%) had TCA. This patient segment (with complications) represents 34% of those with long colonic aganglionosis and 62% of those with TCA in the study sample. Catto-Smith et al[17] reported that a substantial proportion of the interviewees limited their physical or social activity because of soiling or odor. Patients with a history of long-segment disease had overnight soiling more frequently than those with short segment disease (often/ always: long segment 9/ 18; short segment 8/53; $P < 0.01$). Soiling stool was more lose in consistency (long segment 12/ 13; short segment 10/13; $p< 0.05$).This difference could be explained by the following findings. First, our sample size of the study is small. Second, all of our patients underwent DPT as the definitive surgery, while in the study by Moore et al, different surgical procedures have been done. Also, we have excluded cases of TCA in our study.

The timing of pull-through surgery is a controversial issue and the current trend is to do the pull-through operation early. However, Gunnarsdottir et al[18] could not find any statistical difference in the QoL in the adults who underwent surgery before 6 months of age as compared with those who were operated on later.

The benefit of early diagnosis and treatment is that most of the mid-term complications can be managed well before the child starts going to school, thus preventing any social embarrassment and loss of school days. Most of the pediatric surgeons can perform pull through operation in infants without much difficulty hence one must try to diagnose and treat HD at the earliest. It is worth mentioning that with improved diagnostic availabilities and better access to healthcare for more populations, now children are presenting at a younger age than before. In our study, the youngest patient to have presented was 4 months old. In contrast, a few years ago, the patients used to first present as late as 7-8 years. On comparing BFS among index cases no statistically significant difference was found concerning the age at which the patient first presented with symptoms. Thus, it is concluded that the age at presentation has no impact on the final outcome.

Gunnarsdottir et al[18] found that females had significantly lower mean scores for general health and mental health than the the age and sex-matched controls in Swedish general population. Females had also lower mean scores than males in all subgroups. Women with HD had a lower QoL in adulthood and needed special consideration in their follow-up. Reviewing their medical records did not reveal any plausible explanation.

The unique feature of this study is that unlike most of the previous studies, it is a longitudinal study wherein we have assessed the efficacy of targeted therapy in improving BFS. The studies available in the literature provided only a cross-sectional evaluation of QoL.

There are few demerits of the current study. The cohort studied was retrospective; the operative notes of patients did not specifically mention if it was Martin's modification or otherwise. Small sample size with a limited period of study is another demerit. The relationship between the outcome versus the type of HD and sex of patients needs to be interpreted with caution due to fewer cases of long-segment HD and number of females in the study. It was not possible to assess the outcome of different surgical procedures as DPT is the predominant surgery done at our institute. The BFS questionnaire does not take into account episodes of enterocolitis which is a catastrophic complication of HD. So, its impact on the overall outcome could not be assessed. The study design does not take into account urinary dysfunction which according to some studies is quite prevalent following DPT. This study excluded cases of TCA and HD patients with associated Down syndrome. These patients are generally expected to have more problems related to bowel control. To test

the efficacy of BFS, the study must include these patients as well.

To conclude, this study showed that a significant number of HD patients continue to have bowel problems for many years after definite surgery. Parents or caretakers of patients must be counseled regarding the need of long-term follow up which extends to many years after definitive surgery. These patients face significant gastrointestinal problems (constipation with or without soiling).Using bowel function scores, we can identify patients who need early targeted interventions including medical and surgical therapy. Most patients present with mid-term complications, while few patients have long-term complications, though the functional outcome among both these groups of patients is similar. The residual spur seen in a few patients, suggests that the use of appropriate size staplers and Martin's modification of Duhamel pull-through at initial surgery might avoid this complication. Extent of HD, age at the initial presentation and gender do not affect the final outcome. Short-term targeted therapies for 3-month appears to have favorable results.

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## APPENDIX -1

Box 1: Bowel Function Score Questionnaire

| Factors | Score |
|---|---|
| ***Ability to hold back defecation*** | |
| ❖ Always | 3 |
| ❖ Problems < 1/week | 2 |
| ❖ Weekly problem | 1 |
| ❖ No voluntary control | 0 |
| ***Urge to defecate*** | |
| ❖ Always | 3 |
| ❖ Most of the time | 2 |
| ❖ Uncertain | 1 |
| ❖ Absent | 0 |
| ***Frequency of defecation*** | |
| ❖ Every other day or twice a day | 2 |
| ❖ More often | 1 |
| ❖ Less often | 0 |
| ***Soiling*** | |
| ❖ Never | 3 |
| ❖ Straining<1/week, No change of underwear required | 2 |
| ❖ Frequent staining, change of underwear often required | 1 |
| ❖ Daily soiling requiring protective aids day and night | 0 |
| ***Fecal accidents*** | |
| ❖ Never | 3 |
| ❖ < 1/week | 2 |
| ❖ Weekly requiring protective aids | 1 |
| ❖ Daily requiring protective aids | 0 |
| ***Constipation*** | |
| ❖ None | 3 |
| ❖ Manageable with diet modification | 2 |
| ❖ Manageable with laxatives | 1 |
| ❖ Manageable with enemas | 0 |
| ***Social problems*** | |
| ❖ None | 3 |
| ❖ Sometimes | 2 |
| ❖ Problems restricting social life | 1 |
| ❖ Severe social / psychological problems | 0 |

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Case Report

# Accessory Scrotum: A Note on Nomenclature, Diagnostic Criteria and Classification

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**Keywords**
Accessory scrotum
Scrotal malformation
Perineal hamartoma
Perianal lipoma

**Abbreviation**
LSF - Labio-scrotal fold

**Abstract**

*A newborn with pedunculated perianal lipoma and accessory scrotum is reported. By reviewing the 60 cases of accessory scrotum documented in the literature, we propose a standardized definition, diagnostic criteria and clinico-embryological classification of this rare malformation.*

## CLINICAL DESCRIPTION

A full-term male newborn presented at birth with asymptomatic soft pedunculated mass hanging from the anal margin. The mass was bilobed; one of which was a 2x3 cm soft tissue mass covered with smooth normal colored skin, while the other lobe was an empty sac of dark pigmented wrinkled skin resembling that of scrotum.(Fig 1) There were no other congenital anomalies as revealed by clinical examination and screening ultrasonography. The mass was completely excised under caudal analgesia. Post-operative recovery and healing was uneventful.

Histologically, the two lobes differed significantly; one of which showed smooth muscle bundles (dartos) dispersed in dermal collagen (Fig 2) while the other lobe showed abundant mature adipose tissue in deep dermis.

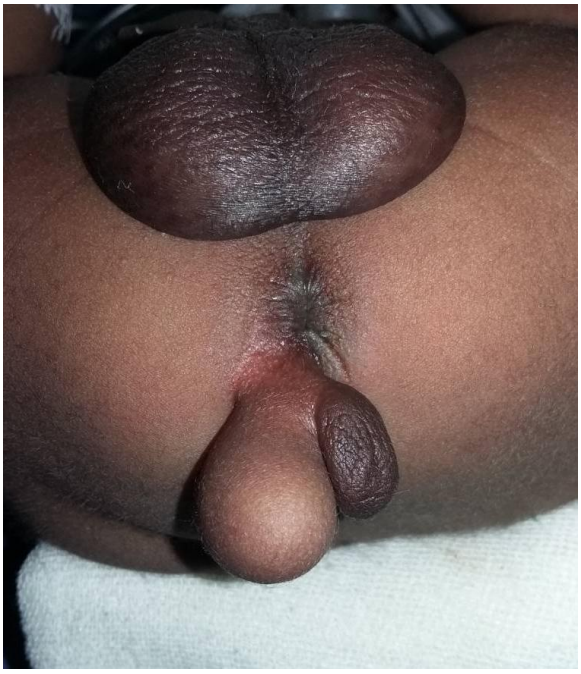

**Fig 1.** *Clinical photograph showing pedunculated peri-anal mass. The bilobed mass shows lipomatous and scrotal components. Normal scrotum is also seen in addition to accessory scrotum.*

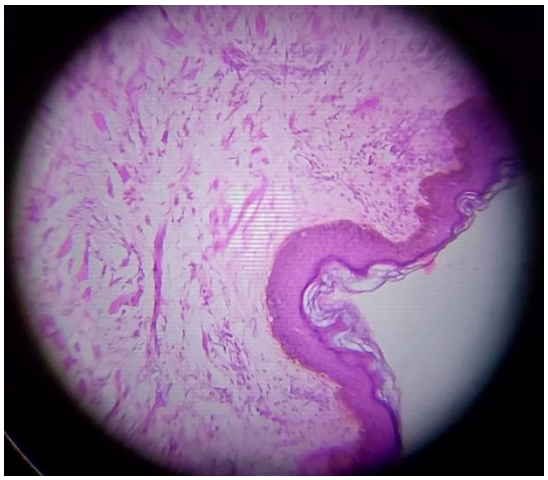

**Fig 2.** *Histopathology of the accessory scrotum showing skin layers with dartos muscle underneath. (Hematoxylin and Eosin staining; 100X magnification)*

## DISCUSSION

Accessory scrotum is an extremely rare malformation with fewer than 60 cases been recorded in the literature.[1,2] It is defined as ectopically located supernumerary scrotal sac sans testis. It is usually located posterior to the normally formed orthotopic scrotum (e.g. perineum or perianal margin).[3] Only 3 cases of accessory scrotum at other locations (e.g. groin, pubis and penile shaft) have been described.[4] Nearly 80% of them are associated with lipomatous perineal hamartoma.[2,5]

'Ectopic scrotum' is yet another rare anomaly that is defined as ectopically located hemi-scrotum containing ipsilateral testis.[3] Absence of hemi scrotum at orthotopic location is characteristic of this condition. Unlike accessory scrotum, they are located anterior to native scrotum (e.g. groin, pubis, upper thigh)[6] and are not associated with lipomatous hamartoma.

As ectopic location is common to both, 'accessory scrota' have been misreported as 'ectopic scrota' and vice versa in the literature.[7] Diagnostic labeling of an abnormal perineal tissue as 'accessory scrotum' may sometimes be difficult if the skin wrinkles are less prominent or if it is just a tiny skin tag. To avoid confusion in nomenclature and clinical labeling, we propose a set of diagnostic criteria that distinguishes 'accessory scrotum' from 'ectopic scrotum' and other hamartomatous lesions. (Box 1)

High incidence of coexisting perineal tumors (71% perineal lipoma, 6% lipoblastoma, 3% mixed hamartoma) in accessory scrotum is thought to be of embryopathogenic significance.[8] Sule proposed that lipomatous overgrowth (hamar-toma) of caudal mesenchyme in a 4-12-week-old embryo might disrupt the continuity of developing labioscrotal folds (LSF) which are destined to become future scrotum.[9] The detached LSF might then develop into accessory scrotum, while the native LSF continues to develop into normal orthotopic scrotum. Such mechanical disruption is also possible in lower limb malformations wherein fetal heel presses upon the developing LSF.[10]

Interestingly, none of the accessory scrota, that are associated with perineal lipoma or tibial malformations, had coexisting anomalies. In contrast to this, accessory scrota without perineal

Box 1: Diagnostic criteria of accessory scrotum

| |
|---|
| **Major criteria** |
| 1. Presence of normal bipartite orthotopic scrotum in addition to ectopic scrotal tissue |
| 2. Absence of testicular tissue (macroscopic or microscopic) within the ectopic scrotal sac |
| **Minor criteria** |
| 1. Ectopic tissue with wrinkled dark pigmented skin macroscopically resembling a scrotum |
| 2. Ectopic tissue with concentration of androgen receptors similar to that of scrotum |
| 3. Ectopic tissue being located posterior to the orthotopic scrotum |
| 4. Histological demonstration of dartos smooth muscle within the aberrant tissue |
| 5. Bipartite / bilobed morphology of ectopic tissue analogous to that of normal scrotum |
| Diagnosis of accessory scrotum necessitates **both** major criteria **plus** any two of the minor criteria. |

lipoma often had serious malformations of genitourinary system, anus and other internal organs. Thus, there seems to be three different embyo-pathgenic types of accessory scrota:

**Type 1:** Typical accessory scrotum associated with perineal tumors (e.g. Perineal lipoma, hamartoma, lipoblastoma) or lower limb deformities (e.g. Tibial aplasia). They are probably due to mechanical disruption of the developing labio-scrotal fold. (Sule's theory)[9]

**Type 2:** Typical accessory scrotum not associated with perineal mass. They are probably due to defective regional organizer cells of embryonic perineum. The resultant field defects may include malformations of caudal end derivative such as genitourinary tract and anorectum. Any one of the following mechanism may play a role in its embryogenesis: (i) Aborted attempt of caudal duplication (Lamm-Kaplan's theory)[7], (ii) triple primordial anlage of the labioscrotal swelling (Takayasu's theory)[11], (iii) Abnormal division of LSF and posterior migration of the detached segment (Coupris-Bondonny's theory)[12]

**Type 3:** Atypical accessory scrotum that occurs as a part of syndromic genetic mutations. Distant malformations such as cleft lip and heart disease are common in this type and the accessory tissue may be located anterior to native scrotum.

Several attempts have been made to classify accessory scrota. Park[6] classified associated lipoma into protruding (sessile) or peduncular. He opined that the former type supports Sule's theory while the latter one supports Takayasu's theory. Ratan[13] classified it into 3 types: Type A - accessory scrotum presenting as a tag of rugose skin over a lipoma; Type B - accessory scrotum presenting as a well developed sac; Type C - accessory scrotum associated with pseudo-duplication of external geintalia. Hoar[14] classified it based on anatomical location such as supra-inguinal, femoral, penoscrotal and perineal. He found that femoral type was more associated with distant anomalies like cleft lip; suprainguinal type was more associated with other genitourinary anomalies while perineal type wass an isolated malformation. Amman[15] classified the clinical morphology of accessory scrotal tissue into 3 types: ill defined (faint) rugacity, tiny skin tag (or nodule) and well developed rugose skin. Kumoro[16] classified it into mid-perineum type (satisfying Takayasu's theory) and the lateral type (satisfying Lamm-Kaplan's theory). These classifications lack practical significance, while our classification is clinically applicable. For example, in accessory scrota associated with perineal lipoma (type-1) elaborate investigations are not necessary to rule out coexisting anomalies, while type-2 requires imaging studies of genitourinary tract and type-3 requires detailed genetic work-up.

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Clinical Update

# Biliary Atresia

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**Keywords**
Biliary atresia
Jaundice

**Abbreviations**
**BA**- Biliary atresia
**KPE**-Kasai Portoenterostomy
**GGT**-Gammaglutamyl transferase
**ICL** - Intrahepatic cystic lesions
**LT**-Liver transplant

**Abstract**

*Biliary atresia is a common cause of neonatal jaundice. This article reviews the recent literature on biliary atresia. Recently near-infrared fluorescence cholangiography (NIR-FCG) using indocyanine green fluorescence (ICG) has been employed during Kasai portoenterostomy (KPE) to identify bile flow from hilar ductules. Intrahepatic cystic lesion (ICL) is a newly recognized complication of KPE. Patients with ICL developing after six months of KPE tend to have better outcomes. Usually, ICL are preceded by episodes of cholangitis. Higher GGT levels before KPE is now identified as a risk factor of post-operative cholangitis.*

## INTRODUCTION

Biliary atresia (BA) is a neonatal disease which presents as progressive inflammatory obliteration of the extra-hepatic ducts. This condition is the most common cause of persistent direct hyperbilirubinemia during the first three months of life. Furthermore, it is also the most common indication for liver transplants in children.

BA affects 1 in 15,000 live births and patients are mostly females. Without treatment, BA may cause cirrhosis, liver failure, and death. Even though its exact cause remains unknown, some possible etiologies have been suggested. Among them, transplacentally acquired viral infections were found to have strong association. Up to 68% of the affected infants have serum antibodies against reovirus type-3. Additionally, some genetic mutations have been identified as potential causes of developmental anomalies. Cases of polysplenia, malrotation, situs inversus, pre-duodenal portal vein, and absent inferior vena cava are some examples of developmental malformations associated with BA.

BA can be classified according to the localization of the biliary obstruction. (Fig. 1) Biliary atresia type-I (5%) represents obliteration of the common bile duct. Type-II (2%) can be subdivided into IIa and IIb (according to the Kasai classification) and it signifies obliteration of the common hepatic duct. In type-IIa, the cystic duct and common bile duct remain patent; thus, atresia is limited to the common hepatic duct. In type-IIb the common hepatic duct, cystic and common bile duct are affected. Type 3 is the most common form (more than 90% of cases) in which ductal obliteration extends to common bile ducts, cystic

duct, and left and right main hepatic ducts at the level of the hepatic transverse fissure.

Among treatments for BA, Kasai portoenterostomy (KPE) and liver transplant are known as leading therapies. KPE may be regarded as a primary surgical approach, but liver transplant is ultimately required for most of the patients. Prognosis can be determined by the promptness of work-up and referral to surgery. Degree of liver fibrosis and ductal plate malformations have also been related to prognosis.[1-4]

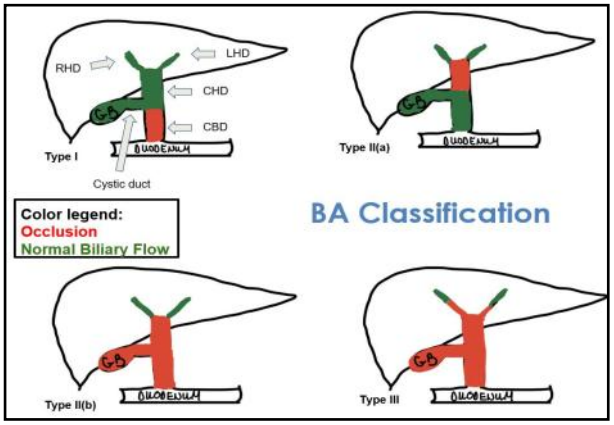


**Fig 1.** *Classifications of Biliary Atresia. RHD-right hepatic duct; LHD-left hepatic duct; CHD-common hepatic duct; CBD-common bile duct; GB-gallbladder. Orange colour - atretic segment, green colour-patent duct*

## CLINICAL FEATURES

The clinical characteristics for BA include insidious jaundice by the second week of life. Upon evaluation, vital signs typically appear to be normal. Hence, despite apparently healthy look the infant may progressively develop acholic stools, choluria, and hepato-splenomegaly. Although not specific for this disease, weight loss and loss of appetite may be observed. Icterus may be noted at birth and it may persist for more than three weeks. Jaundice and acholic stools in an infant are specific but not sensitive indicators of BA. Cholestatic jaundice of infancy caused by neonatal hepatitis and BA are very similar. Thus, identification of BA may sometimes be complex and several differential diagnoses should be considered. (Box 1)

## INVESTIGATIONS

Laboratory investigations used to identify BA include elevated total bilirubin level in serum with 50-80% of conjugated bilirubin. Lipoprotein-X levels are also elevated (> 300 mg/dl). Gamma-glutamyl-transpeptidase (GGT) level may be greater than 200 units/dl. Furthermore, lab tests utilized to exclude BA may include perinatal infectious panel (TORCH titers, hepatitis profile), indicators of metabolic disorders ($\alpha$-1-antitrypsin levels) and markers of hemolysis (Coomb's test, reticulocyte count, peripheral smear).

GGT is an important enzyme measured in BA. It is an epithelial transferase associated to glutathione and its conjugate. Elevated levels of this enzyme indicate cholestatic diseases and it accurately differentiates BA from other cases of neonatal cholestasis. High GGT levels (> 500 IU/L) have been associated to lower rates of native liver survival. Yet, GGT levels are not routinely tested for and more research is needed to assess its prognostic worth.[5,6]

**Box 1. Differential diagnosis of Biliary Atresia**

- Physiologic Jaundice
- TORCH Infection
- Idiopathic Neonatal Hepatitis
- $\alpha$-1-Antitrypsin Deficiency
- Alagille Syndrome
- Biliary Hypoplasia
- TPN-Induced Cholestasis.

## IMAGING

In terms of imaging, ultrasound has been considered as the initial diagnostic tool of choice. It reveals triangular cord sign, absent gallbladders, and intra- or extra-hepatic bile ducts and liver parenchymal echogenicity. Demonstration of post-prandial contraction of the gall bladder eliminates the possibility of BA, further enhancing its diagnostic relevance. Length and contractility of the gall bladder is also suggested to aid diagnosis.

However, several studies have concluded that the triangular cord sign is more useful sign rather than length and contractility of the gallbladder.[7]

Hepatobiliary scintigraphy, also known as Di-isopropyl imino-di-acetic acid (DISIDA) scans, is the diagnostic imaging of choice. These tests allow for evaluation of hepatic uptake and bilio-enteric excretion. The presence of this radio-isotope in the patient's gastrointestinal system excludes BA as a final diagnosis.

## INVASIVE STUDIES

The three invasive studies are considered in the diagnosis of BA are percutaneous biopsy, laparoscopy, and mini-laparotomy. Firstly, the percutaneous biopsy was regarded as a safe procedure with a complication rate lower than 1%. It is deemed as a highly specific and sensitive method. Although these are not specific for BA, percutaneous biopsy allows evaluation for liver damage, ductular proliferation, bile plugs, giant cell transformations and fibrosis. (Fig. 2) However, immuno-histochemistry of the portal ducts may demonstrate the presence of the epithelial membrane antigen in large ducts, which is a specific feature of BA.[8-16] A histological scoring system has been developed at the Children's Hospital of Fundan University. This 21-point system demonstrated good diagnostic accuracy and consisted of eight features: liver fibrosis, portal ductal proliferation, bile plugs in portal ductules, cholestasis, hepatocellular changes inflammatory cells infiltration in portal region, extramedullary hematopoiesis, and ductal plate malformation.[20]

Nonetheless, it will be important to point out that preoperative biopsies are not indicated when laparoscopies are available. Biopsies are often considered as an unnecessary postponement delaying KPE.[17] On the other hand, combination of the laparoscopic technique along with additional tests may serve to diagnose BA accurately. This route spares the liver from trauma and provides a safer diagnostic protocol.

Alternatively, the mini-laparotomy is regarded as a final diagnostic alternative. It is majorly used for gall bladder cholangiograms and liver biopsies. Small hypoplastic ducts are associated to Alagille syndrome.[13] In BA, the gallbladder will be atrophic fibrous remnant or when present will be filled with white bile and no communication to biliary tree or distal extra-hepatic communication.

## SURGICAL APPROACHES

Symptoms of BA are due to the inability of the system to excrete conjugated bile and inflammatory obliteration of bile ducts. Therefore, delaying without treatment may lead to an increase in fibrosis and a decrease in ductal size. These sequence results in a poorer prognosis. Conversely, surgery before sixty days may lead to improved prognosis and bile drainage in 75-80% of the cases. Thus, patients should be assessed early and referred promptly to surgery.

Preoperative management involves supplemental formula feed with medium chain triglycerides and fat soluble vitamins (A, E, D, and K). Parental education and support are recommended as well.

Operative management for KPE involves administration of intravenous fluids and prophylactic antibiotics. Surgeon's expertise is also crucial for proper technique during the procedure. In a given case, identification of severe cirrhosis should call for cessation of KPE and the patient should be referred for liver transplantation. The steps for KPE include beginning with a mini-laparotomy to rule out all other differential diagnoses. When BA is confirmed, the incision is extended and KPE is performed. Some studies have pointed out the importance of KPE as a temporizing procedure of future liver transplant surgery. Using a low subcostal incision well below the costal margin is essential for incorporation during a future liver transplant.[14] Moreover, this procedure consists of removing the obliterated extra-hepatic biliary system and making a jejunal conduit for bile drainage. The proximal jejunum is

attached to the jejunal conduit through a Roux-en-Y anastomosis. Extensive dissection of the duodenum and right colon should always be avoided and usage of a long jejunal loop for conduit reconstruction is recommended.[18-23]

Although feasible, a laparoscopic approach to KPE is shown to be unfavorable when compared to postulated advantages of laparoscopy.[4] Recently near-infrared fluorescence cholangiography (NIR-FCG) using indocyanine green fluorescence (ICG) has been employed during KPE. This technique facilitates observation of hilar micro-bile ducts and provides real-time visualization of bile flow during KPE. When compared to control groups, without NIR-FCG groups showed higher rate of postoperative normalization of hyperbilirubinemia.[22]

Post-operative management of KPE requires monitoring of liver function, bile flow, cholangitis and portal hypertension. Prevention of further complications must remain a top priority; thus, nutritional and familial support should always be available. Early and recurrent cholangitis lowers survival chances.[10] Cholangitis should be treated aggressively with antibiotics. Higher GGT levels before KPE is a risk factor for post-operative cholangitis. Occurrence of intrahepatic cystic lesions (ICL) is another potential complication post-KPE. Patients with ICL developing after six months of KPE tend to have better outcomes. Usually, ICL are preceded by episodes of cholangitis. Clinical symptoms for ICL may include fever, jaundice, leukocytosis, and acholic stools. Solitary cysts are amenable to percutaneous drainage. Portal hypertension may manifest as esophageal varices, hypersplenism, and ascites. Sudden cessation of bile flow, malabsorption, and pruritus are other complications of KPE.

When KPE fails, a liver transplant (LT) is needed for most patients. Total bilirubin levels greater than 2 mg/dl and albumin levels lower than 3.5 g/dl at three months of KPE are predictive of the necessity of LT. In fact, BA is the most common cause for pediatric LT. KPE remains as the initial surgical treatment choice for BA but is only considered a temporary solution. Minimal supra-colic dissections during KPE reduce post-LT morbidity and mortality. Bowel perforations following LT in BA are unfavorable. All in all, BA patients have excellent long-term survival chances.[18]

## CONCLUSION

Although relatively rare, BA remains as the most common cause of end-stage liver disease and LT. Persistent cholestasis in newborns must be assessed with urgency. In this sense, KPE should be offered to infants before their sixty days of life. One-third of BA patients become long-term survivors with KPE. LT is indispensable for patients with failed KPE, liver failure or late referral to surgery. Some common indications for LT include bilirubin levels above 10 mg%, low albumin, weight loss, and uncontrolled ascites.

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