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Pediatric Surgery in Tropics

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Pediatric Surgery in Tropics
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Editorial

Balancing the Priorities

Naeem Khan

Veteran Pediatric Surgeon, Islamabad, Pakistan.

Change is a constant phenomenon. The changes that I have seen in the last 65 years of my medical career, and as to how they have influenced my personality, are interesting to look back. My initial years of student life were mostly non-serious, dedicated only to passing examinations and becoming a doctor, with no clear objectives. But coming in contact with seniors and elders during the house job and seeing suffering patients-and-families, changed the direction of my life. Initially, I was in the junior orthopedic job - not by choice but by what was available - followed by travels for earnings and a desire to specialize. I ended up in England in 1965, where again a massive change of atmosphere influenced my direction, which has lingered on till today.

First and foremost was that during duty hours, the punctuality, discipline and sense of responsibility that I saw in doctors, nursing staff and the para-medical staff was beyond description. Secondly number of patients seen in outpatient department, was always by pre-arranged appointments. This was very different from our country, where patients by the hundreds turn up as outpatients. With such an orderly arrangement, detailed history-taking and in-depth clinical examinations were possible. This allowed us to reach the most plausible clinical diagnosis and directed us to request appropriate investigations rather than a snap decision of ordering ultrasonography, CT scan or MRI on the patient's first utterance of "abdominal pain". A known fact to everyone is that overcrowding and having to see sometimes more than 300 hundred patients in an out-patient department of a public hospital is no mean a task. It is certainly counterproductive to our health services.

Let me briefly move on to Tripoli, Libya where there was monitory affluence; but a lack of punctuality matching that of our continent. In other words, I am alluding to three aspects which are essential for the progress of our health service system, namely discipline, punctuality and financial availability.

For an equitable provision of health service to the entire nation, especially to the poor and the illiterate, a system should be evolved where we should be able to serve all with long-established surgical procedures, keeping in mind the peculiar issues such as our financial means, overcrowding of hospitals and most importantly, the availability of medical and paramedical staff. We should refrain from selling our skills of giving small or no scar in places where availability of equipment is non-existent or procurement of spare parts and maintenance is a difficult task. Till such time of parallel progress in manufacturing our own medical equipment and its repairs, we must seriously design our own strategy of delivering health services.

Let me submit that private set-ups may continue to provide the state-of-the-art minimal access

procedures to the affluent and the insured people; but as long as understaffed, overcrowded, ill-equipped public hospitals exist, the standard expedient procedures serving the majority of the population should remain in place. Medical education in our medical schools and the surgical skills of young doctors, even if it is at the cost of a scar, should remain in line with our prevailing circumstances and it should be our major goal. In our materialistic environment, one should be cautious of salesmen and manufacturer's inducements which they come up with to sell expensive equipment like robotics. In such circumstances, we should wisely consider the institutional interests and the needs of patients.

Finally, I would like to submit that a skillful surgeon is recognized not only from his dexterity but also from humane approach and kindness towards patients.

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Editorial

Jugaad: Lessons from the Tropics to the World

Ketaki Gharpure

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Jugaad is here to stay. The Oxford English Dictionary defines 'Jugaad' as "a flexible approach to any problem-solving that innovatively uses limited resources". In very simple terms, it stands for 'put together'. The word finds its origin from Northern India where farmers used essentially spare parts to assemble their own vehicles known as 'Jugaad'. Gen-Z talks about DIY (Do it yourself) and hacks; the millennials (Gen-Y) have lateral and out-of-the-box thinking. However, the decades-old concept of jugaad encompasses exactly that - the approach to finding solutions with improvisation and ingenuity.

A search on PubMed on Jugaad yields just about 8 results all published in the last decade. Prakash *et.al* [1] break the concept of Jugaad into 4 stages:

- Yearning for the solution of a problem
- Learning and experimentation
- Exploring available options and resources
- Designing a creative solution

To summarize, this is paraphrasing of the old adage, "Necessity is the mother of inventions".

The main concern around this quick-fix approach in healthcare is safety and legality. However, if the jugaad approach is taken, it is fairly straightforward to innovate using existing tools and work with manufacturers to develop low-cost solutions. This low-cost solution must meet the benchmark of safety and cross the hurdles of cost and access.

Every surgeon in training or practice, faces a problem that they solve with what is essentially a jugaad. These small ideas are seldom deemed worthy of sharing or publishing; but therein lies a hidden untapped cache of innovations waiting to see the light of day.

Jugaad is not a new concept in pediatric surgery. Almost every instrument, device and equipment began with utility in adult practice which is subsequently adapted for pediatric use. Pediatric surgeons a couple of decades ago had to innovate and find smaller-sized alternatives, all through what was jugaad in its essence. To cite a few examples, Ramakrishna *et.al* [2] have published their idea of innovating a 5mm trocar using the trocar of a suction-drain (Romovac™). Kulkarni *et.al* [3] have described a jugaad for jet insufflator in difficult airway intubations. These are just examples of papers with 'jugaad' in the title; there are be many more without that word. The author remembers an innovation in which a feeding tube within an endotracheal tube and wall suction was used to create an effective Replogle suction tube and many similar handy innovations not formally published. There is value in highlighting ideas such as these.

Pediatric Surgery in Tropics should, therefore, encourage all pediatric surgeons to write about their 'jugaad' ideas. If a particular jugaad is reproducible and resonates with readers, many

might put it into practice. Even if it is not implementable, it will at least give the readers an impetus towards problem-solving approach.

Today, even the so-called resource-abundant healthcare systems are facing difficulties, as the price of healthcare continues to skyrocket. Healthcare systems across the world are crumbling under the pressures of juggling the economic burden of providing healthcare. Therefore, cost-cutting through innovation is not something to be looked-down upon; rather it is something to be legitimized with due process towards safety. *Jugaad* is nothing new but, it is now time for the world to employ this concept to use what little is available, to accomplish a lot, or to use everything that is available with utmost efficiency.

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Clinical Study

Phlebectasia and Phlebothrombosis of the Penile Veins in Children after Non-Medical Circumcision

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Keywords
Male Circumcision
Complications of Surgery
Penile Phlebectasia
Penile Vein malformations

Abbreviations
DPV - Dorsal Penile Vein
MC - Male circumcision
PMD - Penile Mondor's Disease
VM - Venous malformations

Abstract

Background: *Abnormally enlarged, tortuous and dysplastic penile venous vasculature (phlebectasia) are rare in children. The prevalence of this anomaly and its possible etiologic correlation with circumcision is examined in this study.*

Patients and methods: *This is a prospective comparative cohort study of 830 children, aged 6 months to 12 years, enrolled between June 2021 and December 2023. Among them, 420 were circumcised (group A) and 410 were intact (group B). All of them were physically examined and investigated with Doppler ultrasound for any abnormality of the penile venous vasculature. The two groups were compared for any difference in the incidence of different anomalies of the penile veins and penile deviation or chordee. Data were analyzed by Mann–Whitney U-test and a P-value less than 0.05 was considered significant.*

Results: *Phlebectasia (21/420) and phlebothrombosis (3/420) were more frequently seen in the circumcised group, but they were rare in the intact group (2/410). These venous abnormalities were commonly seen in the dorsum of the penis (15/24). In circumcised patients, the phlebectasia was associated with penile deviation to the left side in 13 cases and a ventral penile chordee in 1 case. But, in uncircumcised children, the prominent dorsal penile vein was not associated with any chordee. An associated dilation of the deep dorsal penile vein was confirmed in 15 of the 24 cases.*

Conclusion: *Circumcision wound and the subsequent scar may increase the incidence of abnormal penile venous vasculature which is commonly associated with penile deviation.*

INTRODUCTION

Unlike many other veins of the human body, penile venous drainage is specially designed. The usual pattern of 'veins outnumbering arteries' is reversed in the penis. In contrast to paired dorsal arteries, dorsal penile veins (DPV) are unpaired and single which are arranged in deep and superficial layers. Superficial DPV starts distally and it mainly drains the skin of the penis and prepuce. This vein, as it courses proximally in the midline within the subcutaneous tissue, receives numerous tributaries from the skin of the penile shaft. Deep DPV lies between the Buck's fascia and the tunica albuginea. It receives drainage from the distal two-thirds of the corpora cavernosa via emissary veins and the corpus spongiosum via circumflex veins.[1] The superficial DPV ultimately drains into the left and right superficial external pudendal veins, a tributary of the great saphenous vein. Deep DPV drains into the prostatic plexus. Recently, another small pair of dorsal veins has been found that lie just deep to the deep dorsal vein, but above the tunica albuginea.[2] The unique venous anatomy is intended to facilitate penile erection; the engorged corporal tissue compresses penile veins and venules to maintain penile erection.

Variations in the appearance of the superficial veins of the penis are most innocuous and do not require any medical attention. Several factors like age, ethnicity, tumescent state, genital pigmentation and hormonal effects influence the clinical visibility of penile veins. 'Veiny penis' (penile phlebectasia) is the term used to refer to the normally visible, dilated, painless superficial veins of the penis. It is commonly seen in elderly fair-skinned men. On the other hand, penile varicose veins are similar to phlebectasia but the veins are torturous. Penile vein thrombosis is yet another painful condition with visibly engorged veins. All the 3 entities closely mimic each other.

In phlebectasia the swollen and twisted veins may occur due to inherent weakness of the venous wall or due to increased pressure within the veins. Although phlebectasia may not pose any serious medical risk, it can cause discomfort during erection and is unsightly.

Male circumcision (MC) is associated with a wide spectrum of complications, which may be early or late. However, abnormal penile venous vasculature as a complication of MC was not reported before. The association between post-circumcision abnormal DPV and penile deviation (lateral chordee) has also not been studied. It appears to be secondary to the distorted elastic penile tissue and altered anatomy that follows MC.[3] This paper is intended to examine this unusual complication of MC.

PATIENTS AND METHODS

Eight hundred and thirty healthy children with normal penile size were included in the study. Children who had major systemic diseases or intellectual disabilities were excluded. Baby boys below 3 months of age or those above 12 years and those with any un-related penile anomalies like microphallus or webbed penis were also excluded.

This prospective cohort study was approved by the ethical committee for human experimentation at the authors' universities. Also, this study was carried out following the ethical standards of the hospital's institutional ethics committee on human experimentation. Parents of all study subjects signed a written informed consent for photographing their children and they also consented to the usage of the photographs and data of their children for publication, but with masked identities.

A total of 830 children were examined prospectively between June 2021 and December 2023. They are divided into two groups: Group A

consisted of 420 children aged 4 months to 12 years who underwent non-medical MC. They were evaluated at 2 to 5 months (mean 3 months) after non-medical MC. Group B consisted of 410 age-matched intact (uncircumcised) children, who sought medical help for unrelated conditions like hernia or hydrocele, but without any detectable congenital anomalies of the penis.

Diagnosing superficial phlebectasia of the penis is mainly clinical. The examination commenced sequentially for the circumcision scar, coronal sulcus, frenular remnants, penile shaft, glans, and urinary meatus. Assessment of the visibly dilated veins was done in both flaccid and erect states. In those who required a surgical operation, erection was induced artificially by intra-corporal injection of saline. Any hardness or tenderness elicited was also recorded. The length and the diameter of visible veins were measured by a digital caliper (Fixtec™, Model FHVC0151) with a resolution of 0.01 to 0.1 mm. The degree of penile deviation was measured according to the Sarkis-Sadasivam method[4] by using a sterile protractor. Deviation of the meatus from the midline after aligning the protractor with the penile shaft was taken as the reference point for measuring the penile deviation.

Dilation of the DPV was assessed by colored Doppler ultrasound. Primary hyper-coagulability was excluded by investigating the bleeding and coagulation profiles. For all circum-cised children, the age at the time of MC, the technique of operation, the duration of bandage and any detect-able bleeding or other early compli-cations were recorded. For those with early post-MC bleeding, the method of hemostasis was also noted.

In children with dilated DPV who deserved surgical intervention (n=21; Group A) the procedure was done through a circumferential or semi-circumcision incision. Partial penile degloving (n=8), or more extensive degloving maneuver (n=13) was done to expose serpiginous tortuous superficial DPV which is sandwiched between the skin and the Buck's fascia. The fibrotic tissue of the dysplastic Buck's fascia (Fig.1) that is respon-sible for lateral deviation was excised and penile alignment was restored with absorbable sutures. The deep dorsal veins were inspected for any dilatation or varicosity, and its recognition was enhanced by an opening made on the Buck's fascia and the corpora cavernosa was milked analogous to the squeezing of a balloon.[5] The superficial DPV was stripped thoroughly and ligated with 6-0 nylon suture. The remaining healthy Buck's fascia (recognized as fleshy sheet of fascia) was approxi-mated using undyed 6-0 Vicryl™ sutures. (Fig.1) All the patients were operated under general anesthesia as day-case. The postoperative course of those who underwent ligation of superficial DPV was uneventful except for minimal lymphatic edema of the preputial remnant in 2 cases and it resolved spontaneously.

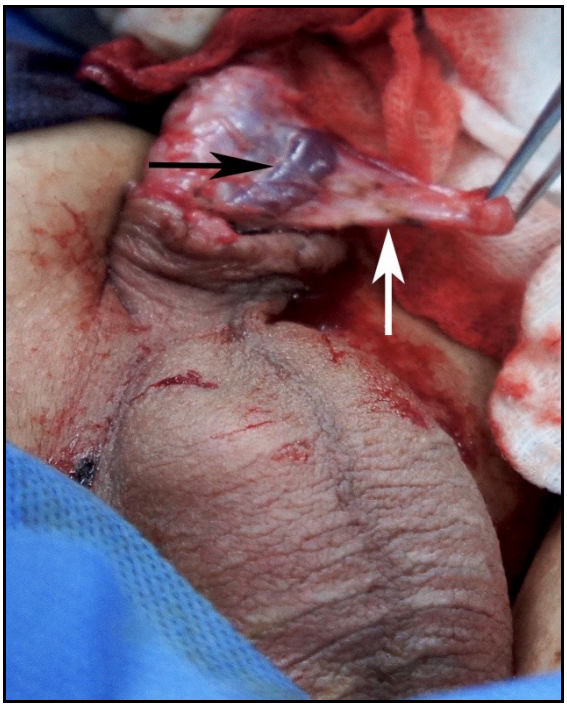

Fig 1. *Prominent dilated dorsal vein (black arrow) along with dissected dysplastic Buck's fascia (white arrow). (NB: The dorsal penile vein appearing on the ventral side is a photographic illusion caused by the rotation of the penile shaft during operative mani-pulation.)*

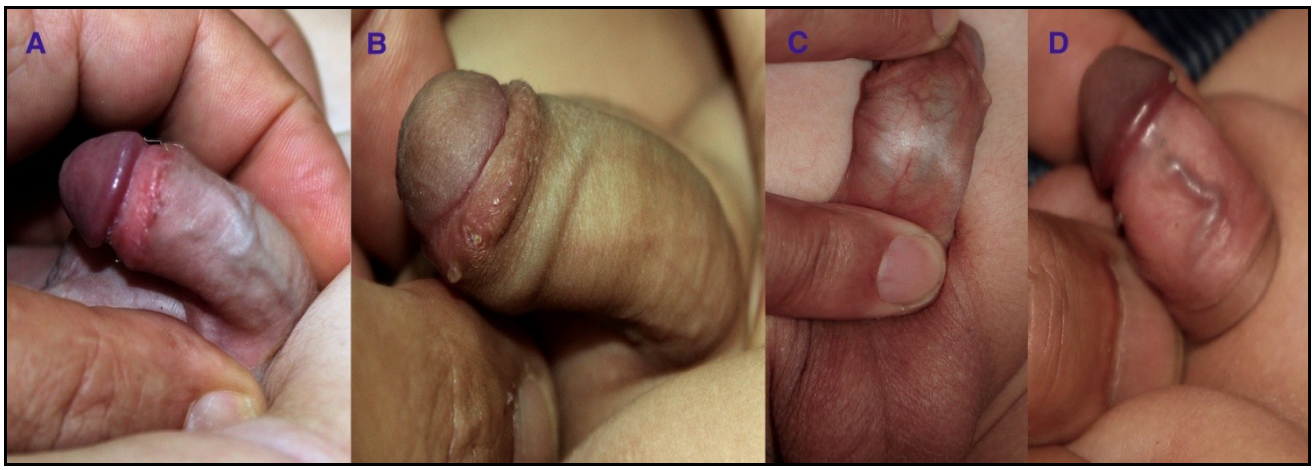

Fig 2. *(A) Prominent dorsal penile vein (DPV) in a circumcised 2-year-old boy; (B) Transversely abnormal DPV giving the appearance of a double coronal sulcus; (C) A case of ventral prominent veins; (D) Prominent vein confined only to the inner preputial layer (retracted view)*

The venous status and any residual deviation were assessed before skin closure by artificial erection induced by using saline infusion into the corporal bodies. Finally, the wound was approximated while an assistant surgeon consistently stretched the skin of the penile shaft.

Cases diagnosed with phlebothrombosis (n=3; Group A) were managed conservatively for 2 weeks and they had uneventful outcomes. Two cases diagnosed in the control group (n=2; Group B) were simply followed up by reassuring the parents without any therapeutic intervention. The follow-up period of those with abnormally dilated DPV in both groups ranged from 6 to 22 months (mean 7.7± 4.9 months; n = 26).

Discrete data were analyzed by Mann–Whitney U-test. Data with continuous values are expressed as mean ± standard deviation. Student's paired t-test was applied for inferential purposes after logarithmic transformation. Statistical significance was set at $P < 0.05$.

RESULTS

In 16 patients of the group-A, the superficial DPV was distended longitudinally in the midline for about 2cm in length (range 18- 32 mm) reaching the preputial remnant or only confined to the inner preputial layer. Transversely abnormal DPV was rare (n=3; Group A) and it gave an appearance of a 'double coronal sulcus'. Ventral phlebectasia was seen in 2 patients (Fig 2). The overlying skin was tethered and occasionally erythematous.

Penile deviation to the left side was diagnosed in 13 cases and ventral chordee in one case. (Fig 3) An associated suprapubic depression (deficiency of the suspensory ligament) was noted in 3 cases (Fig 3C). Penile phlebothrombosis with a hard, cord-like, thrombosed vein was felt in 3 patients; one of them also had a bluish discoloration of the skin around the dilated vein; in one case it was painful on palpation. (Fig 4)

Detailed history of MC procedure revealed that all the 24 cases of the Group-A were circumcised during the first month of life and the prepuce in all of them was cut by guillotine method using a bone-cutting forceps. Nine cases had thermal cauterization for hemostasis and in 4 cases a monopolar diathermy was used. Post-MC bandage was left *in situ* for 2 days in 12 cases. Post-MC hematoma developed in 4 cases. Post-procedure infections occurring in 5 cases were managed with

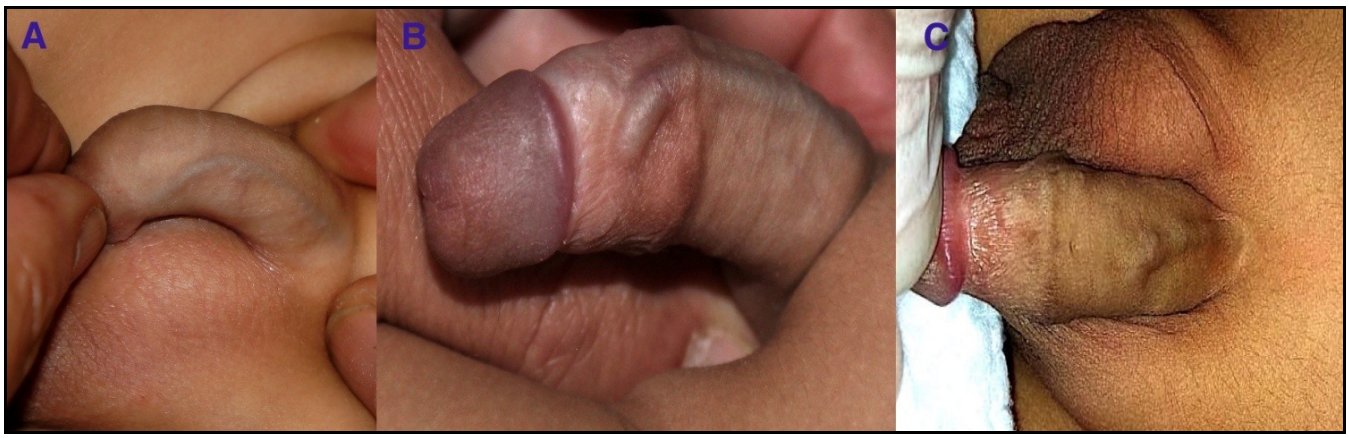

Fig 3. *(A) Prominent dorsal penile vein (DPV) with chordee; (B) Prominent DPV with a left sided penile deviation; (C) Prominent DPV with a suprapubic depression.*

systemic antibiotics and local ointment. Phlebectasia was recognized by the family after a mean period of 3 months after circumcision (range 2-5 months). All the patients of both groups had normal coagulation profiles. Dilation of the deep DPV was confirmed in 15 of the 24 cases with dilated super-ficial DPV in the group A; but in none of group B.

Statistical analysis showed a significant correlation between circumcision status and abnormally prominent penile veins. Frequency of phlebectasia (5% vs 0.5%; P<0.001), penile deviation (3.57% vs 0%; P<0.004) and phlebothrombosis (0.71% vs 0%; P<0.256) were all more frequent with group-A than with group-B. (Table 1)

DISCUSSION

Visibly dilated penile veins are not rare in adults as they are seen in 24% of individuals.[6] However, abnormally dilated, tortuous, serpiginous penile veins in children are rare and are scantly reported in the literature. Prominently visible penile veins may be due to phlebectasia, varicosity of the DPV or thrombophlebitis (Mondor's disease of the penis). Acquired phlebothrombosis of the DPV may be due to distorted local anatomy as in circumcision, coagulation disorder or disorders of hemoconcentration (e.g. leukemia). Sometimes it is considered as sequelae of hamartomatous embryonic remnants [7] and it may also complicate penile surgeries like circumcision.

Penile phlebectasia appears as asymptomatic, abnormal dilatation and enlargement of the penile veins. When complicated by phlebothrombosis it appears either as a bluish raised and serpiginous tract or as a bluish soft nodule. They may be single or multiple, and the lesions usually measure 0.5 to 1 cm in diameter. The thrombosed area may be tender. The dilated vein usually disappears under slight pressure but refills spontaneously.

Table 1: Comparison of venous abnormalities of the penis

Variables	Group A* (n=420)	Group B* (n=410)	*P* - value
Dilated DPV	24 (5.5)	2 (0.5)	<0.001
Longitudinal DPV	16 (3.8)	2 (0.5)	0.002
Transverse DPV	3 (0.7)	0 (0.0)	0.256
Dilated VPV	2 (0.5)	0 (0.0)	0.490
Phlebothrombosis	3 (0.7)	0 (0.0)	0.256
Lateral chordee	13 (3.1)	1 (0.2)	0.004
Ventral chordee	1 (0.2)	0 (0.0)	1.000

** Values in parenthesis are percentage*
DPV - Dorsal penile vein; VPV - Ventral penile vein

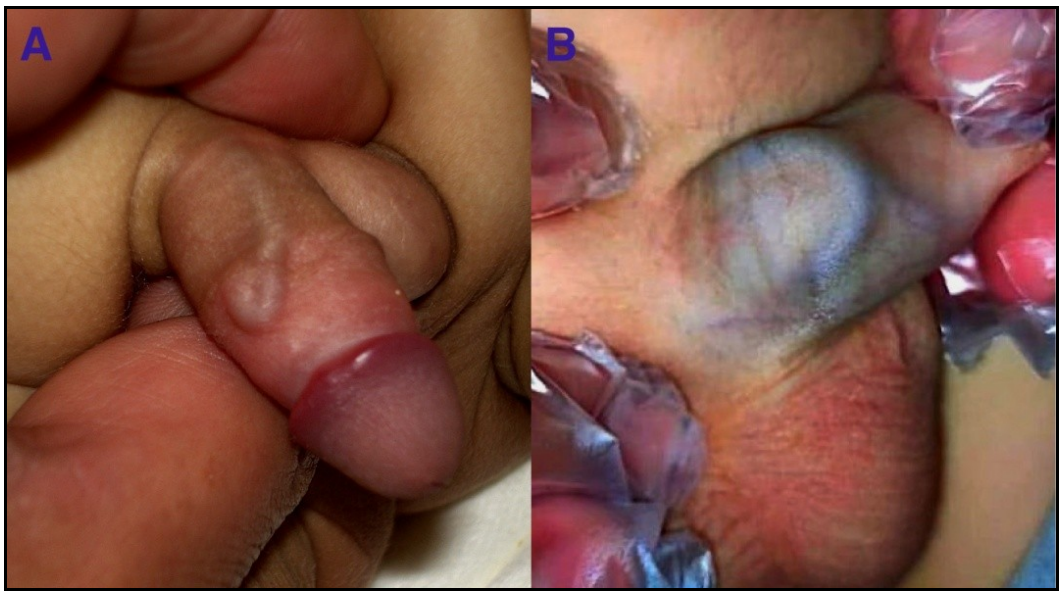

Fig 4. *(A) Prominent dorsal vein in penile phlebothrombosis; (B) Bluish discoloration associated with thrombosis.*

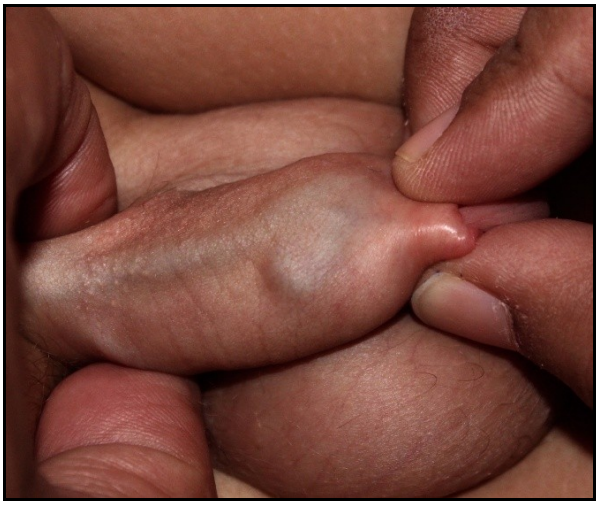

Fig 5. *Dialed tortuous vein in an uncircumcised child*

Phlebectasia is common in adults and the elderly, possibly secondary to hormonal and physiologic stimuli.[8] It has no significant functional impact on erection or intercourse; but may cause problems with body image and cosmesis. Rarely superadded vascular occlusion by thrombosis may result in erythema, bruising or painful swelling. Minor bleeding into the subcutaneous plane may also complicate these lesions.

Penile phlebectasia should be differentiated from rarities like varicose vein of the penis, penile hemangioma, and penile Mondor's disease (PMD). PMD is a rare condition characterized by thrombosis of the superficial DPV.[9] It is common in adolescents and is often underdiagnosed. It is characterized by sudden onset; hard, painful, rope like palpable structure on the penile dorsum; and is well appreciable during erection. Penile color duplex ultrasound is an important tool in diagnosing and monitoring this condition. Lack of blood flow and non-compressibility of the dorsal vein indicate thrombosis.[9] The PMD is a self-limiting condition. However, anticoagulants such as low-molecular-weight heparin can be used to treat thrombophlebitis. Persistent thrombus may rarely remain visible as a hyper-echoic linear shadow on the dorsum of the penis.[10]

Penile hemangioma may be superficial, deep or combined. They may be in proliferating or involuting phase. Infantile hemangiomas are not inherited but may be formed secondary to mutation in a primitive stem cell which is responsible for the developing blood vessels.[11]

Venous malformations (VM) are the commonest vascular anomalies and can be found anywhere in the body, but those of the external genitalia are quite rare. Although they may be small, they may cause significant psychological impact and functional impairment. Management of penile hemangioma is usually challenging.[12] Owing to rarity, treatment has not been standardized.[13] Usually VM slowly worsen over time and never regress spontaneously. They clinically appear as bluish soft vascular masses.[14] Most of the VM are often asymptomatic. Sclerotherapy and laser have become viable options for treating small lesions. Although penile VM often relapses or demands multiple treatments, the prognosis is generally favorable. Percutaneous sclerotherapy of penile VM in children under the guidance of digital subtraction angiography is safe and effective, without affecting the cosmetic appearance and function of the penis.[15]

Proper clinical examination and history can differentiate abnormally prominent DPV from the other lesions. In this study, penile phlebectasia is characteristically more common on the dorsal aspect and runs longitudinally in the midline of the dorsum of the penis reaching to the preputial remnant of circumcised individuals. Transversely dilated DPV are rare and give the appearance of a double coronal sulcus; ventral dilated veins are extremely rare. Although clinical appearance is enough for diagnosis, colored Doppler ultrasonography may be used for precise understanding of the anatomical and physiological features. Rarely, direct phlebography may be indicated, to confirm the clinical diagnosis. For a more comprehensive study of the venous system, magnetic resonance angiography (MRA) is beneficial, and it may be particularly helpful in PMD.[16] However, in our cases, Doppler ultrasound was sufficient for the detection of both the dilated superficial DPV and any anomalies that may affect the deep venous vasculature.

Penile curvature (lateral chordee) and torsion are generally not rare. Its prevalence is 0.5 to 10% during adolescence or young adulthood.[17] The exact etiology of this deviation is not well known, but it may be genetic. Pathogenic mechanism of lateral chordee or torsion includes abnormal skin and dartos fascia attachment, asymmetry of the tunica albuginea of the corpora cavernosa, abnormal bony fixation of the corporal bodies or dysplastic Buck's fascia.[17]

Male circumcision (MC) is usually practiced for several reasons, such as social, religious, cultural, or rarely for medical indications like phimosis. This unnecessary procedure is largely performed for secular reasons in the Western world, which is sustained by a variety of rationalizations such as aesthetic value. Although MC is considered as one of the oldest and the most common surgical procedures practiced globally, recently the rate of MC has been declining across several countries.[18] This declining rate may reflect the changes in demographic patterns and parental beliefs raised by studies in psychology and ontogeny.

In this prospective comparative cohort study of 24 cases of pediatric penile phlebectasia were diagnosed 2-3 months after neonatal MC was done for non-medical reasons. There is significant correlation between MC and the occurrence of penile phlebectasia. Families reported that there had been no such abnormal veins noticed before MC. Even the physician who performed the MC had not noted any pre-operative phlebectasia. Although this could be a subjective correlation, but the high frequency of phlebectasia in circumcision group as compared to the control group is highly significant. New onset penile torsion in 13 cases and chordee in 1 patient in association with abnormal veins in 69% of children is yet another point that supports our hypothesis.

We propose that post-circumcision bleeding and the use of aggressive hemostatic techniques may have a pathogenic link to the onset of post-MC

phlebectasia by distorting the Buck's fascia. Histological comparison of the penile scars shows extensive fibrosis, abundant amounts of collagen and absence or paucity of normal smooth muscle cells more often with using electro-coagulation than with using ligatures.[19] None of the patients in this series had diabetes, hormonal insufficiency or other sort of trauma except circumcision.

Venous stripping was superior to venous ligation in terms of cosmetic look and it also entailed a careful relief of fibrotic tissues.

Thus far, there have been no scientific studies that support regeneration of penile veins following their surgical excision. Therefore, not only the surgical outcome is promising, but also morphologic complications are minimal.[3] When post-circumcision bleeding occurs, it is safer to apply compression or applying fine stitches than using any mode of electro-coagulation. This is essential to avoid any deleterious effect on the penile vasculature; to preserve erectile capability and to avoid infection especially, in the delicate penile tissue of neonates.[19] Early detection and management of penile phlebectasia may have a positive impact on affected children not only in terms of general psychological wellbeing but also on their sexual life. Abnormal penile vasculature may contribute to male sexual dysfunction.[20]

CONCLUSION

Post-MC penile phlebectasia with or without phlebothrombosis is infrequently reported. Its actual incidence could be greater than what is accounted. Due to the embarrassing nature of genital symptoms, patients may not be reporting to physicians of dilated veins. Distorted Buck's fascia following circumcision could be the common etiology of both penile phlebectasia and penile curvature.

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Clinical Study

Hypospadias Repair: A Single-Centre Experience with Clinical Presentations, Operative Techniques and Outcome

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Keywords
Hypospadias
Koyanagi operation
Penile malformation
Thiersch- Duplay repair
Urethro-cutaneous fistula
Urethroplasty

Abbreviations
TIPU - Tubularized Incised Plate Urethroplasty

Abstract

Background: *Hypospadias is the most common congenital abnormality of the penis. Its incidence is on the rise; however, its reconstructive repair remains a huge challenge. We reviewed our experience with an aim to expand our understanding and modify the treatment protocols.*

Patients and Methods*: This is a retrospective evaluation of all consecutive cases of hypospadias repaired at a Nigerian Teaching Hospital between January 2013 and June 2018. Data on demography, presentations, type of anomaly, repair techniques and outcome were extracted from case records and analyzed.*

Results*: A total of 64 boys had primary repair for hypospadias. The median age at presentation and at repair was 17 and 28.5 months respectively. At presentation, 32% had already been circumcised and 71% of them had been done by nurses. The most commonly associated anomaly was inguinal hernias in 3 (5%) and chordee in 39(61%). The location of the meatus was glanular in 2 (3%), coronal in 20 (31%), distal penile in 17 (27%), mid-penile in 6 (9%), proximal penile in 5 (8%), penoscrotal in 7 (11%), interscrotal in 2 (3%) and perineal in 5 (8%). Case volume doubled in two successive years with a peak of 15 per year. Techniques of repair were tubularized incised plate urethroplasty (TIPU) in 51 (80%), Koyanagi in 7 (11%), Thiersch-Duplay in 4 (6%) and staged repair in 2 (3%). Urethro-cutaneous fistulae occurred in 16 (25%); however, in 12 (75%) fistula healed spontaneously. There was a drop in fistula rate from 40% in 2015 to 6.3% in 2018.*

Conclusion*: The incidence of hypospadias in our center is on the increase. In proximal variants staged repairs or Koyanagi technique have better outcomes than TIPU. Sustained practice and modification of techniques reduce complications.*

INTRODUCTION

Hypospadias is the most common congenital penile abnormalities with an incidence of 1 in 250 males in the United States of America.[1,2,3] Its incidence is found to be on the increase in Western countries and this has been attributed to disruption of endocrine milieu by environmental pollution.[2,4] There is paucity of epidemiological data from Africa; however, Okeke et.al[5] reported an incidence of 1.1% in a community study from south-east Nigeria.

According to the location of the external urethral meatus, hypospadias is classified into anterior (glanular, coronal and subcoronal), middle (distal penile, midpenile) and proximal (proximal penile, penoscrotal, inter-scrotal and perineal).[6] Hadidi further sub-classified the anomalies for standardization and comparison of various subtypes.[7] He introduced factors such as the nature of the prepuce, the urethral plate, presence or absence of chordee, penile rotation and scrotal transposition. The most commonly associated anomalies are cryptorchidism and inguinal hernias.[8]

Repair of hypospadias continues to pose huge reconstructive challenges to pediatric surgeons, urologists and plastic surgeons. There have been over 300 techniques described in literature;[1,9] yet none have been successfully and satisfactorily applied to all the variants of the anomalies by all surgeons. Each case is repaired on its merits and according to the surgeon's experience.

The pediatric surgery unit of our hospital began routine repairs of hypospadias in 2013. With an increasing caseload, we considered it necessary to review our experience to highlight the unique challenges and overall outcome in our settings.

PATIENTS AND METHODS

This is a retrospective study of all consecutive cases of hypospadias that were repaired at the Nnamdi Azikiwe University Teaching Hospital, Nnewi, Nigeria between January 1, 2013 and June 31, 2018. Data on demography, clinical presentation, type of anomaly, repair techniques and the outcomes were extracted from operation theatre registers, patients' case notes and ward records.

All the patients were seen in the clinics of our two pediatric surgery units. Repairs were done by 3 pediatric surgeons and the choice of technique was dependent on the location of the meatus, status of prior circumcision and the experience of the surgeon. We employed 4 major techniques of repair namely, tubularized incised plate urethroplasty (TIPU), Koyanagi repair, Thiersch-Duplay technique and staged procedures. All the procedures were done using fine instruments and atraumatic polyglactin-910 sutures (size 6-0 and 5-0). Hemostasis for the mid-penile and distal variants was achieved with tourniquet application while that of proximal types was achieved by subdermal infiltration of lignocaine with adrenalin (1:100,000 dilution) along the line of incision. It was injected using a 29G needle mounted on a 1ml syringe. The urethral stents used were either pure silicon catheters or appropriate-sized feeding tubes. The stents were allowed to drain freely into the diapers without connecting them to urobags. Suprapubic cystostomy was not done in any patient for urinary diversion. Antibiotic coverage was given with a third-generation cephalosporin (Ceftazidine). Small-size penises were augmented with 1-3 doses of intramuscular testosterone 25 mg, given 3 weekly prior to surgery. Patients were followed up for 3months -5 years.

Data extracted were entered in a Microsoft Excel spreadsheet and were analyzed using the Statistical Software Package for Social Sciences (IBM SPSS Version 22 Chicago Il). Results are presented in frequencies, percentage (Tables 1-3). Association between categorical variables was tested with the Chi-square test and the statistical significance was set at a p value < 0.05.

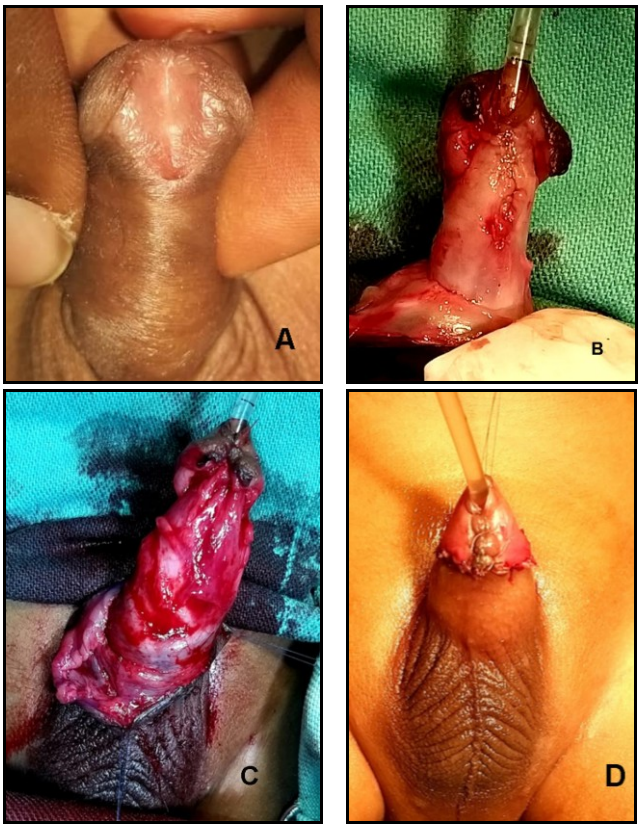

Fig 1. *Modified tubularized incised-plate urethroplasty: (A) Distal penile hypospadias in uncircumcised penis; (B) Penile shaft degloved and neo-urethral tubularization done in 2 layers with polyglactin-910 after vertical midline incision on the urethral plate; (C) Vascularized Dartos fascia cover as third layer; (D) Final appearance after glanuloplasty and penile resurfacing.*

RESULTS

A total of 64 boys had primary repair of hypospadias within the study period. Our case volume per year was 4 in 2013 (6%), 8 in 2014 (13%), 15 in 2015 (23%), 12 in 2016 (19%) and 10 in the first half of 2018 (16%). The median age at presentation was 17 months (range 2 weeks - 9 years) while the median age at repair was 28.5 months (range 6 months -10 years).

Abnormality of the penis was noticed by parents in 44(69%), doctors in 14(21%), nurses in 5(8%) and grandmother in 1(2%). Table-1 summarizes the types of hypospadias encountered based on the location of the meatus. Chordee was present in 39(61%). The more proximal variants are significantly associated with increased frequency of chordee ($P = 0.001$).

Among the 21 boys (33%) who had had circumcised prior to presentation, 15 (71%) were done by nurses, 4 (19%) by doctors, 1 (5%) by traditional circumcisionist and 1(5%) by unknown person.

The most commonly associated co-morbidity was inguinal hernias in 3(5%); others include hemophilia, hydrocele and patent ductus arteriosus (PDA) - each found in 1(2%) patient. Various techniques of repair employed are represented in Table 2. The mean duration of antibiotic therapy was 9.6±2 days (range 5-14).

Complications were reported in 25 (39%) patients (Tables 1-3). Urethro-cutaneous fistula occurred in 16 (25%) however 12(75%) of them healed spontaneously. Of all those who had various complications, only 10 (16%) needed a second surgery. None required a third operation.

Table 1: Frequency of the types of hypospadias and post-operative complications

Location of the meatus	n (%)	Chordee n (%)	Complication n (%)
Glanular	2 (3%)	0 (0%)	1 (50%)
Coronal	20 (31%)	12 (60%)	5 (25%)
Distal penile	17 (27%)	12 (71%)	7 (41%)
Mid-penile	6 (9%)	6 (100%)	3 (50%)
Prox. penile	5 (8%)	5 (100%)	3 (60%)
Penoscrotal	7(11%)	6 (85%)	4 (57%)
Interscrotal	2 (3%)	2 (100%)	1 (50%)
Perineal	5 (8%)	5 (100%)	1(20%)

Prox. - Proximal

Table 2: Technique of hypospadias repair and complications

Surgical Technique	n (%)	Complications n (%)*
TIPU	51 (80%)	20 (39%)
Koyanagi	7 (11%)	3 (42%)
Thiersch-Duplay	4 (6%)	2 (50%)
Staged Repair	2 (3%)	0 (0%)

TIPU - Tubularized incised plate urethroplasty
** P = 0.001*

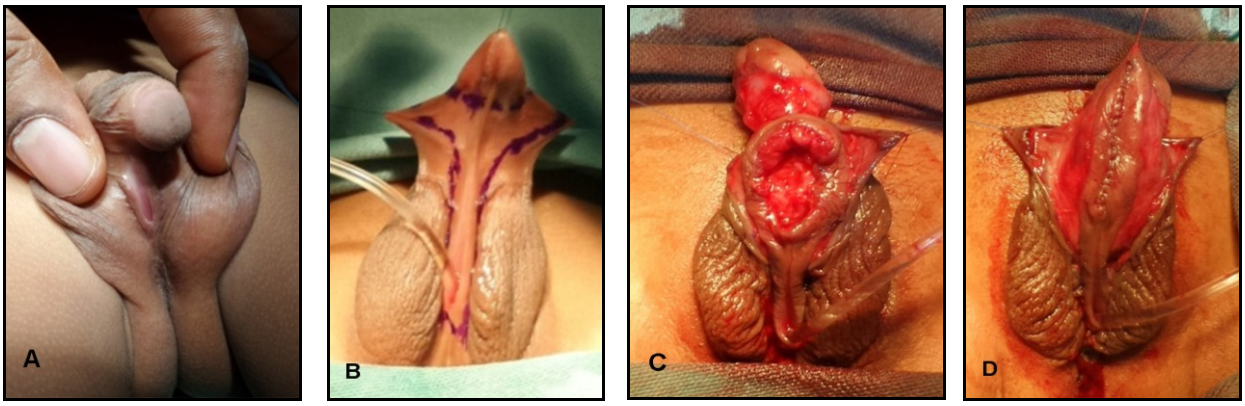

Fig 2. *Single stage repair of perineal hypospadias using Koyanagi technique: (A) Perineal hypospadias with about 40 degrees chordee; (B) Markings of the proposed incision; (C) Transposition of the prepuce ventrally for tubularization; (D) Approximation of the prepucial flaps; (E) Penile resurfacing after neourethral tubularization of the preputial flaps.*

There was no complication in those who had staged repair (P =0.001). There was a drop in fistula rate from 40% in 2015 to 6.3% in 2018.

DISCUSSION

A two-fold increase in the case volume of hypospadias in our practice was observed for the first 3 years after which it reached a plateau before rising again in the first-half of 2018. This experience appears to be in concordance with the reported global trend of rising incidence.[2] It might also have been due to increased referrals to our center since we started providing the services. Possibly our case volume could have increased still further, but for the limited number of operating days.

The median age at repair in our practice is much higher than the widely recommended age of 6-18 months.[1,10] Shadrach[11] and Bello[12] operated at

44.9 months and 2 years of age respectively, which is quite similar to that of ours. This may be due to delayed presentations of patients or attributable to the limited availability of operating slots. The later problem was highlighted by Thomas[13] from Lagos, Nigeria where the median age at repair was 4 years.

Table 3: Frequency of complications in hypospadias repair

Complications	n	%
Isolated Urethro-cutaneous fistula (UCF)	10	40
Partial wound breakdown (WB)	6	24
UCF with meatal stenosis	5	20
UCF with complete wound breakdown	1	4
Bleeding	1	4
Isolated meatal stenosis	1	4
Meatal stenosis with partial WB	1	4
Total	25	100

UCF - Urethro-cutaneous fistula, WB - Wound breakdown

In our series, most of hypospadiac defects were noticed by parents. Although we could not ascertain the place of delivery in this study due to its retrospective nature, this finding underscores the need for a complete evaluation of all newborns by a trained medical person at the time of neonatal circumcisions.

The most common type of hypospadias in our series was anterior hypospadias (61%) followed by the posterior and mid penile types (30% and 9% respectively). This is quite similar to previous studies[8,10,11,14] that are both from within and outside Nigeria. Chordee was present in 39(61%) while absent in 25(39%) patients. Chordee was present in all mid-penile and proximal variants, but none in the glanular types. Frequent association of chordee with more proximal anomalies was statistically significant. Two Nigerian series reported a pattern of chordee identical to that of our experience.[11,15]

In this study, 21(33%) were circumcised prior to initial presentation and 71% of the circumcisions had been performed by nurses and 19% by doctors. This is really of great concern. Although a majority of these anomalies were noticed by the parents, circumcision offered another opportunity of detection by healthcare professionals, but that was missed. There may be a need for re-education and retraining of nurses and doctors on the contraindications of circumcision.[16] Prior circumcision does not affect the outcome of distal hypospadias repairs using TIPU.[17] However, the same may not be true of proximal anomalies and for those who may wish to use other techniques of repair. Hence, we still advocate the preservation of the prepuce in all cases of hypospadias.

We encountered coexisting anomalies in 6 boys (9%) and inguinal hernia was the most common. We did not record any case of undescended testis although it is the most common comorbidity in other series.[11,15]

Four major techniques of repair were employed as shown in table 2. The choice was dictated by the type of anomaly and surgeon's preference. The most commonly utilized technique was the TIPU (80%) and Koyanagi repair (11%). This is similar to the choices of Aisuodionoe-Shadrach et.al.[11] The preference of TIPU over other techniques is attributable to the higher prevalence of distal hypospadias than the proximal variants as TIPU is not appropriate for the latter. In contrast to this, Olajide[15] used TIPU in only 2% of patients. We repaired proximal hypospadias using modified Koyanagi's technique or staged repair. Thiersch-Duplay was used for those distal variants with wide and pliable urethral plates while staged repairs were mostly used in those with proximal hypospadias who were already circumcised. It has been emphasized that no single technique assures consistent success in all variants in the hands all surgeons.[1,18]

All the repairs were stented with per urethral catheter that were allowed to freely drain into diapers. The duration of the stents was between 4-14 days (mean 10.6±2.0). Felicien[18] kept the stents for 15-21 days. Earlier removal of stents may be appropriate in distal hypospadias repairs. In the present study, accidental dislodging of catheter before the 8th postoperative day did not cause increased complications or urine leak. This observation is worthy of persuasion by further research.

Early complications were observed in 25 patients (39%). The complication rate in our study is much higher than that reported by Okoro *et.al* [19] from Norwich (3.6 - 14%); however, they had higher proportion of distal hypospadias repair than us. Our complication rates are similar to that of Abdur-Rahman[20] (33.3%); but higher than that of Bello[12] (20%) and less than that of Thomas[13] (56%). The most common complication was urethro-cutaneous fistula in 25% of patients. Although this was statistically significant, 75% of them closed spontaneously. Overall, only 10 patients (15.6%) needed a second operation which had an overall successful rate of 84%. There were more complications in circumcised children (47.6%) than in uncircumcised (34.9%); but this was not statistically significant (P=0.327). We also observed that perineal hypospadias had fewer complications (20%) than proximal penile hypospadias (60%) (Table 1). We found that 5 out of 7 penoscrotal hypospadias were repaired with TIPU, while all the perineal variants underwent either Koyanagi or staged repair. The Koyanagi technique is considered an alternative to staged repair.[1] Therefore, we believe that proximal hypospadias are better repaired with either staged technique or Koyanagi operation rather than with TIPU. This has also been highlighted previously by Felicien.[18] There was no complication in those who had staged repair. Initially, the overall complication rate in our series was as high as 53% in 2015 before it dropped to 10% in 2018. A drop in complication arte is well known to occur with an increasing case volume.[8]

CONCLUSION

There appears to be an increase in the incidence of hypospadias in our setting. A significant number of them present late and are already circumcised. The major complication of repair remains urethro-cutaneous fistula, most of which closed spontaneously. The rate of complications continuously dropped with growing experience. Finally, the Koyanagi operation of staged procedure gave a better outcome for the more proximal variants.

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Invited Commentary

The Nigerian Experience with Hypospadias Repair

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[2]*Editor, Pediatric Surgery in Tropics.*

In this issue (July-Sep 2024) of the *Pediatric Surgery in Tropics*, Dr. Ugwu *et.al* have reported their experience with hypospadias in Nigerian children.(1) It is commendable that they have improved the results of their hypospadias repairs over a few years. The authors have not mentioned certain details such as the width of the urethral plate, the width of the glans and the degree of chordee following penile degloving. Tubularized incised plate urethroplasty (TIPU) was employed in 51 of the 64 patients, including the 5 out of 7 penoscrotal hypospadias. TIPU is a versatile, reliable, cosmetic and functionally successful surgical method mainly used in distal hypospadias; but in recent times it is also used in some proximal varieties. Its main complications are urethro-cutaneous fistula, urethral meatus stenosis, glans dehiscence and urethral meatus dehiscence that the authors have noted in their series too. The urethro-cutaneous fistula and meatal stenosis rates have been reported as high as 12% and 15% respectively in a recent article.(2) Narrow urethral plate, flat glandular groove and small glans were found to be significantly asso-ciated with higher complication rates in TIPU. Further, Snodgrass advocated that a small glans (usually associated with proximal hypo-spadias) measuring less than 14 mm in width is not ideal for TIPU.(3) Some authors believe that TIPU should not be used if the urethral plate width (UPW) is less than 8 mm.(4,5), though Snodgrass denies the significance of this parameter.(6) Others believe that the functional outcome rather than cosmetic appearance of hypospadias repair is predicted by the UPW.(7)

Mathieu's urethroplasty is a good option for distal hypospadias with a narrow urethral plate and is recommended.(8) A hybrid technique of Mathieu's urethroplasty incorporated with TIPU is also in vogue for distal penile hypospadias with small glans and shallow urethral plate.(9)

Another point of controversy could be the use of a single-stage TIPU in patients with proximal hypo-spadias with significant chordee. There is no mention in the paper about the use of artificial erection test or the use of dorsal plication when single-stage urethroplasty was employed in such patients. Staged procedures should be more liberally performed in such cases as they tend to have better cosmetic and functional results.(10)

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Case Report

Gastrointestinal Bullet Embolism with Spontaneous Expulsion of the Projectile

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Keywords
Ballistic trauma
Gastrointestinal pathology
Bullet embolism
Gastric perforation
Gunshot injury
Intestinal foreign body

Abstract

A 5-year-old girl was admitted with signs of peritonitis following gunshot injury of the abdomen. At laparotomy, a perforation of 1.5 cm diameter on the greater curvature of stomach was noted. Intraoperatively the projectile could not be located and hence gastrorrhaphy was performed. Postoperatively, a plain abdominal radiograph revealed the presence of a projectile in the abdomen. The patient resumed intestinal motility with diarrhea on the postoperative day-4. The bullet was spontaneously expelled during defecation on the postoperative day-7 and the diarrhea resolved on the postoperative day-11. Gastrointestinal bullet embolism with spontaneous expulsion of the bullet is a very rare phenomenon.

INTRODUCTION

Bullet embolism is a rare phenomenon that occurs when a penetrating bullet migrates away from its original entry point through the anatomical channels of the body.[1,2] Most of the cases of bullet embolism are either vascular or gastrointestinal in nature.[2] Cases of bullet migration within the gastrointestinal tract and their spontaneous expulsion through the anus are extremely rare.[3-6] Herein, we report one such case of bullet embolism in a 5-year-old child.

CASE REPORT

A 5-year-old girl was admitted to the surgical emergency department with abdominal gunshot wound from a terrorist attack. On examination, she was conscious, but hemodynamically unstable. Abdominal examination revealed signs of peritonitis. Temperature was 38.3°C, pulse was 146/min

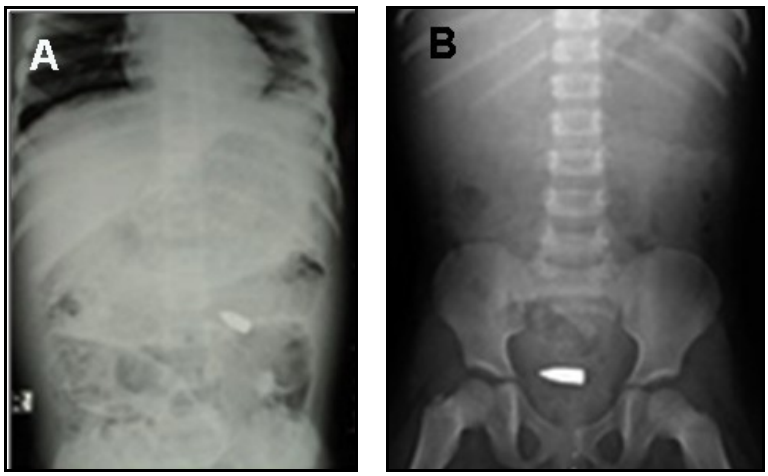

Fig 1. *Projectile located in the left flank on the postoperative day 2 (A) and it migrated to the pelvis on the post-operative day-5 (B)*

and respiratory rate was 56/min. A single, bullet-entry hole over the left anterior chest wall was noticed. The entry wound was oval in shape with a diameter of 1.5cm. There was no hemothorax or pneumothorax. The pre-operative laboratory test revealed hemoglobin of 9.5 g/dl, white blood cells of 9920/mm^3, hematocrit of 28% and platelets of 372 x10^3/mm^3. Plain abdominal radiograph and chest X-ray were not available in the emergency department.

After fluid resuscitation, laparotomy was performed. Intra-operatively a perforation of about 1.5 cm of diameter on the greater curvature of stomach was identified. There was also a splenic injury of Grade II (Moore's classification), without active bleeding. There was no breach of the diaphragm. A search for the projectile was negative. The gastric perforation was repaired.

Patient developed respiratory distress 24 hours after the laparotomy. She had crepitating rales in the left lung with a SaO_2 of 88% on room air. Oxygen was administered. Postoperative investigations revealed a hemoglobin level of 5.9 g/dl and leukocyte count was 12,510/mm^3. On post-operative day-2, a plain abdominal radiograph revealed the presence of a projectile in the abdomen. (Fig.1A) A follow-up radiograph on the post-operative day-5 showed that the bullet had migrated to the pelvis. (Fig.1B)

A chest X-ray showed consolidation of the left lung base. Gentamicin was added to the postoperative mediation. The patient resumed intestinal motility with diarrhea on the postoperative day-4. The bullet was expelled on the postoperative day-7 (Fig.2) and the diarrhea improved on the post-operative day-11. Upon improvement of clinical condition, she was discharged on the postoperative day-17.

Fig.2 *Bullet expelled in stools*

DISCUSSION

In the Western Africa, that is affected by terrorism and civil war, the incidence of abdominal gunshot wounds is high.[7] With the advent of terrorist attacks, daily statistics in our setting shows a significant increase in the incidence of ballistic trauma since 2015. The northern part of Burkina Faso is one of the regions that is most affected by

terrorism. Children are most often the victims of stray bullets.

Spontaneous migration of a retained bullet is a rare phenomenon.[1] This phenomenon occurs when a bullet that penetrates the body is stalled on its pathway and is secondarily carried away from its initial location to a distant site though the luminal structures of the body.[2] The first descriptions of this phenomenon concerned with intravascular bullet embolism was reported by Thomas Davis in 1834.[2] Vascular embolism is the most common form of bullet embolism.[2]

Gastrointestinal bullet embolism is an extremely rare phenomenon, even more rare than vascular bullet embolism.[1] It is rare for a bullet to get stalled in the lumen of gastrointestinal tract.[1] However, a bullet retained in the gastrointestinal tract of survivor may migrate along the lumen, propelled by intestinal peristalsis.[1] On reviewing the literature, only 8 cases of bullet migration through the gastrointestinal tract could be found.[1-6, 8, 9] Among them, there were only 4 cases of spontaneous bullet expulsion during defecation.[3-6] The first case of spontaneous expulsion of a projectile through the anus was reported in 1977 by Weithofer.[3] This was a projectile impacted in the duodenum and the common bile duct for 32 years and was expelled spontaneously during defecation. The case reported by Tebbett [5] is similar to that of our patient. Wani[6] reported an exceptional case of spontaneous bullet expulsion through the anus, in which the gunshot wound was in the chest, and the bullet was lodged near the postero-lateral wall of the trachea. The patient finally coughed up and ingested the bullet before it was expelled in the stool.[6]

In the present case, after penetrating the stomach, the bullet migrated progressively over 7 days within the gastrointestinal tract before being expelled in stools. Intestinal peristalsis might have facilitated the migration and expulsion of the bullet.[1] In addition, handling of the gut during laparotomy may also have caused the migration.[5] The paralytic ileus due to acute generalized peritonitis certainly prolonged the expulsion time.

In the background of hemodynamic instability, signs of infection and features of peritonitis, an emergency laparotomy was performed to assess the lesions. This is fully justified in the absence of sophisticated imaging facilities.

The physical appearance of the projectile (Fig.2) and the damage caused to the body indicate that it must have been shot from a high-velocity weapon from a distant point. The particularity of this trauma is that the bullet shot from a far distance, having passed through the tissues of different densities, will be unstable. This will limit the resultant tissue damage. After penetrating the stomach wall, the projectile, no longer possessing sufficient energy, must have been lodged in the pyloric lumen. As the projectile is contaminated, it could have resulted in enteritis and diarrhea. Although rare, pediatric surgeons must be aware of bullet embolism. A simple X-ray may be sufficient to follow the migration of the projectile.

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CHIEF EDITOR'S COMMENTS

It is a big surprise to know that there are resource-limited settings where even simple imaging facilities such as x-rays are not available for an emergency diagnosis, and surgeons have to work without this basic investigation. We must applaud the initiative taken by the authors from Burkina Faso that resulted in good outcome of the patient.

PST PEDIATRIC SURGERY IN TROPICS

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Case Report

Bile Duct Injury Following Open Cholecystectomy in a Child: A Rare Complication

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Keywords
Cholelithiasis
Bile duct injury
Open cholecystectomy
Gall stones
Surgical complication

Abbreviations
BDI - Bile duct injury
CBD - Common bile duct
CHD - Common hepatic duct, **IHBRD** - Intrahepatic biliary radicle dilatation, **LC** - laparoscopic cholecystectomy, **LFT** - Liver function test, **OC** - Open cholecystectomy, **PTBD** - Percutaneous trans-hepatic biliary drainage

Abstract

Cholelithiasis is an uncommon pathology in the pediatric age group, with an incidence of 0.13-1.9%. The standard of treatment is laparoscopic cholecystectomy (LC). Open cholecystectomy (OC) has been rarely performed nowadays. We report a 5-year-old boy who underwent OC for gallstones and had sustained bile duct injury (BDI), which was missed intra-operatively. The child presented with fever, abdominal collection, and sepsis. We discuss the management of a patient with BDI along with the review of pertinent literature.

INTRODUCTION

Cholelithiasis is uncommon in the pediatric age group, with an incidence of 0.13-1.9 %.(1) Although complications are less common in open cholecystectomy (OC) than in laparoscopic cholecystectomy (LC), the LC is the preferred approach. Bile duct injury (BDI) is a well-reported major complication of cholecystectomy in adults with high morbidity and mortality. The incidence of BDI is 0.04-0.2% in OC and 0.01-1% in LC.(2) Its management includes endoscopic procedures like stenting, sphincterotomy, and percutaneous trans-hepatic biliary drainage (PTBD), which may be either additional or definitive interventions of BDI. There is a lack of literature on BDI management in the pediatric population. In this paper, we are discussing a case of BDI in a child after OC and its treatment.

CASE REPORT

A 5-year-old male child presented at our institute with jaundice, abdomen distension, and deranged liver function test (LFT) following an OC done two weeks prior at another hospital.

As per the records of the previous hospital, he had had cholelithiasis and had experienced symptoms consistent with acute cholecystitis, (viz. pain in the right hypochondrium, fever, and vomiting). Ultrasonography of the abdomen had revealed a

distended gallbladder with multiple calculi, the largest being 11mm located near the neck of the gall bladder. There was also minimal free fluid, indicative of acute inflammation. Surgical intervention had been planned after the failure of conservative management (nil by mouth, nasogastric aspiration, and intravenous antibiotics and intravenous fluids). Due to the unavailability of laparoscopy, an open cholecystectomy (OC) had been performed to address the critical condition (acute cholecystitis). Intraoperatively, the gall bladder was found to be tense, enlarged and was at the risk of perforation. Adjacent structures were severely inflammed and the omentum was adherent to the gallbladder and porta hepatis. This underscores the challenges posed by the absence of LC facilities and the need for timely intervention to prevent complications like gallbladder perforation. In the immediate postoperative period, the child had been apparently stable and was discharged after starting oral feeds. After one week, the child developed fever and progressive jaundice, which had been managed conservatively initially and then he was referred to our centre for further management.

On arrival at our hospital, the child was sick, jaundiced (total bilirubin-17.1mg/dl, direct bilirubin-13.6mg/dl), febrile, and his abdomen was distended with hepatomegaly of 3cm below the costal margin. Ultrasonography of the abdomen showed a loculated collection in the GB fossa, with intra-hepatic biliary radicle dilatation (IHBRD). Magnetic resonance cholangio-pancreatography (MRCP) revealed hepatomegaly with a fluid collection of 9x7cm in the sub-capsular location of segment V of the liver.(Fig.1A) Bilateral IHBRD (Left>Right) with abrupt narrowing and cut-off sign in the common hepatic duct (CHD) was demonstrated. The right anterior duct communicated with the left duct while the right-posterior duct was coursing towards the primary confluence with narrowing at the communication site; the CHD and the CBD were not seen (Bismuth type 3 BDI).(Fig.1B) The ultrasound-guided abdominal drain and PTBD catheter were placed. After receiving two weeks of antibiotics, his clinical condition improved, and LFT normalized. Contrast study done through PTBD tube (Fig.1C), showed opacification of the left and the right biliary radicles; however, there was no contrast in the CHD and the CBD.

The child was then taken up for surgery. On exploration, stomach, duodenum, small bowel loops and surrounding structures were densely adherent to the porta hepatis. It was challenging to delineate anatomy. On flushing saline through the PTBD catheter, the area of leakage was identified, and thus the hepatic ducts were traced. Three ducts, namely the right anterior, right posterior and left hepatic duct, were traced, and the PTBD catheter was seen protruding from the left hepatic duct. A standard retro-colic end-to-side hepatico-jejunostomy was done by single layer anastomosis. The postoperative period was uneventful, and the child was allowed orally on postoperative day-5 and discharged on the day-8. At a 2-year follow-up, the child is doing well with normal LFT.

DISCUSSION

Cholelithiasis is uncommon in the pediatric age group. Although OC is not currently a standard procedure for cholelithiasis, it has fewer complications than LC. The OC gives a better view of the anatomy and thus has less chance of injury to the surrounding structures. In studies comparing LC and OC, LC is associated with more operative time, less hospital stay, and less postoperative pain.[3] Studies have shown that BDI is not because of the surgeon's inexperience but due to the visual misperception of the anatomy in around 70-90% of cases.[4] The frequent causes of BDI include poor anatomical delineation, non-identification of the Calot's triangle, congenital variations, complicated cholelithiasis (e.g. perforation, fibrosis, adhesions, malignancy), difficult hemostasis, technical inexperience and retrograde cholecystectomy.[5]

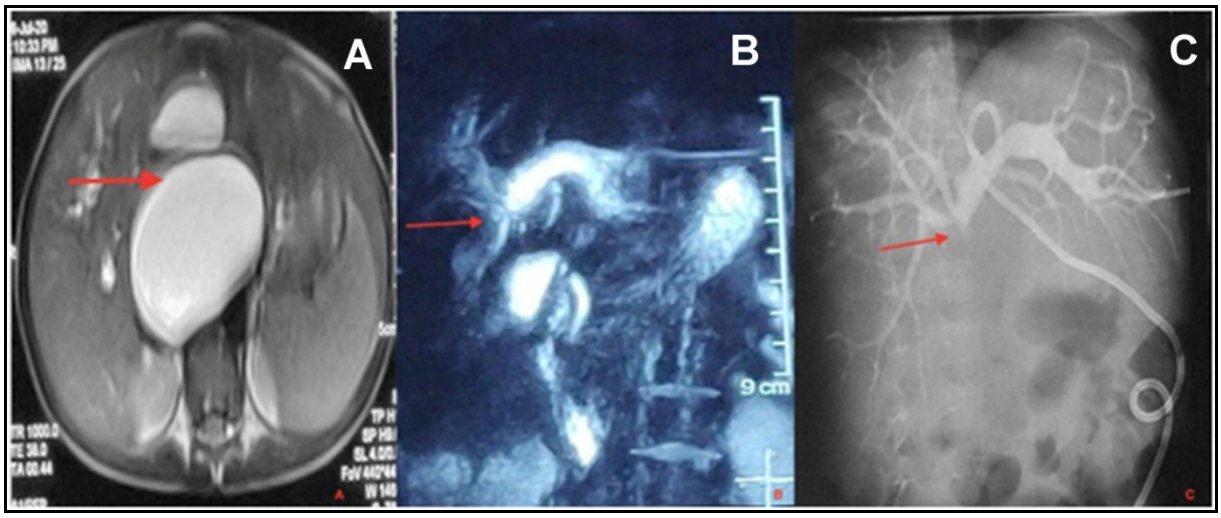

Fig.1 *(A) T2 weighted axial magnetic resonance image showing a collection (arrow) along the surface of the liver. (B) Coronal MRCP (magnetic resonance cholangio-pancreatography) image showing dilated right and left biliary radicles (left > right) with abrupt cut-off (arrow) just beyond the primary confluence. (C) PTC (Percutaneous trans-hepatic cholangiogram) showing the opacification of dilated right and left biliary radicles (left > right) with no passage of contrast across the primary confluence (red arrow) into the common hepatic duct and the common bile duct.*

In the index case, the exact cause could not be identified as the child was operated on elsewhere; the intra-operative difficulties, adhesions, and deviation in bile duct anatomy might be the contributory factors. BDI may lead to bile leakage, intra-abdominal abscess, cholangitis, and secondary biliary cirrhosis.[6]

Once BDI is suspected, on-table intra-operative cholangiography should be done to identify the level of injury and anomalous biliary ducts. Primary repair of the bile duct is ideal, as it prevents morbidity and mortality. If primary repair is not feasible, inserting a catheter into the duct and T-tube drainage will help control local sepsis and will assist in identifying the anatomy during the definitive procedure. The duodenum must be mobilized adequately to attain tension-free end-to-end anastomosis of bile ducts. Multiple studies support that Roux-en-Y hepatico-jejunostomy has the best long-term results.[7] However, in many instances, the injury is not recognized ontable and it presents later with a bile leak as it is in our case. The more delayed the presentation, the more difficult it is to manage because of the inflammation and fibrosis.

The aim of treatment in delayed presentation is immediate management of sepsis, biliary fistula and CBD obstruction. In our case, there was a complete transection of the CBD with injury to the right and the left hepatic ducts. The abdominal drain was placed to decrease biliary sepsis; at the same time, PTBD catheter was inserted to relieve the biliary obstruction. After stabilization elective surgery was done. This management is similar to the algorithm suggested by Andreas.[8]

BDI has been classified by many, and the commonly used classification is that of Bismuth-Strasberg. Depending on the type of injury, the management can be endoscopic, percutaneous or surgical. In general, any fluid collection should be drained.[4] An endoscopic retrograde cholangio-pancreatography (ERCP) is done to evaluate the biliary tree and stenting of the injured duct. PTBD

can be done to drain any collection and provide an alternate channel for bile drainage. It is a low-risk procedure.[9] It decreases sepsis and allows the biliary tract to heal. In case of minor leaks, PTBD alone is sufficient. The injured bile ducts may heal spontaneously or develop stricture, which will require definitive surgery after the critical period is overcome.[6] PTBD catheter also helps in identifying the injured bile duct by injecting either radiographic contrast, fluorescene dye or saline intraoperatively, as we did. The timing of intervention is crucial. In severely septic patients, PTBD alleviates intra-abdominal sepsis and aids in preparing patients for open surgery in case of a major leak, as it is in our case.[6] However, cholangitis can be a severe complication of PTBD; fortunately, the index patient did not have this complication. In transection or stricture of the bile duct, surgical intervention depends upon the availability of bile duct length and its diameter. End-to-end anastomosis of the bile duct has a high chance of leak as is usually under tension. So, a Roux-en-Y hepatico-jejunostomy is preferred.[10]

Prevention of BDI during cholecystectomy is crucial. Identifying the 'critical view of safety' before ligating anything is vital in LC.[11] The infundibular technique and antegrade dissection close to the gall bladder could prevent BDI.[12] Calabro advocated fluorescent cholangiography using indocyanine green.[13] By using infra-red imaging, entire biliary system can appropriately be delineated and hence the anatomical variations, thus decrease the chance of BDI in complex cases.

Managing pediatric BDI is indeed challenging, particularly due to the increased risk of rapid onset of biliary sepsis. Anatomical variations further complicate the scenario, as the small caliber of the duct may not be adequately visualized in preoperative imaging. The insertion of stents or pigtails becomes challenging due to the small size of the ducts, and pediatric-sized equipment may not be readily available in all places. Additionally, addressing BDI in children involves risks associated with interventions such as airway management. Sedation is often required even for investigation, adding another layer of complexity. All these factors underscore the need for a careful and comprehensive approach to pediatric BDI. The management requires a multi-disciplinary approach, and surgery should only be considered after the patient has been stabilized and the ductal injury has been correctly classified.

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Case Report

Meconium Periorchitis in Infants: Role of Conservative Management in Various Clinical Presentations

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Keywords
Acute hydrocele
Meconium disorder
Meconium peritonitis
Peri-orchitis
Scrotal calcification
Testicular swelling

Abbreviations
MP - Meconium peritonitis
MPO - Meconium periorchitis
USG - Ultrasonography

Abstract

Meconium periorchitis (MPO) is an uncommon disease secondary to meconium peritonitis. We report two infants with MPO one of whom presented with acute hydrocele while the other was asymptomatic (incidental diagnosis). Both of them did not have any other associated malformations. Abdomino-pelvic plain X-ray and ultrasound was useful in confirming the diagnosis. Both the patients were managed without surgery. There were no complications at 12-months and 2-months of follow-up respectively.

INTRODUCTION

Between the third month of gestation and birth, the processus vaginalis remains patent, thus establishing a communication between the peritoneal cavity and the tunica vaginalis testis.(1) Therefore, any cause of meconium peritonitis (MP) (e.g. intestinal atresia, intestinal volvulus, cystic fibrosis, or mesenteric infarction) will allow the spilled meconium to reach the tunica vaginalis on one or both sides, causing meconium periorchitis (MPO).(2) It is an uncommon condition, rarely reported in African children.(3)

The diagnosis of MPO is based on clinical suspicion. The circumstances of its discovery vary from accidental diagnosis to acute scrotum mimicking testicular torsion. It may be isolated or associated with other congenital malformations.(3) History, scrotal palpation and imaging studies lead to correct diagnosis. Management of MPO is usually conservative, unless it is associated with other surgically correctable anomalies. However, lack of awareness of this entity may lead to unnecessary surgery.(3) In this communication we share our experience with two cases of MPO that were conservatively managed in our department.

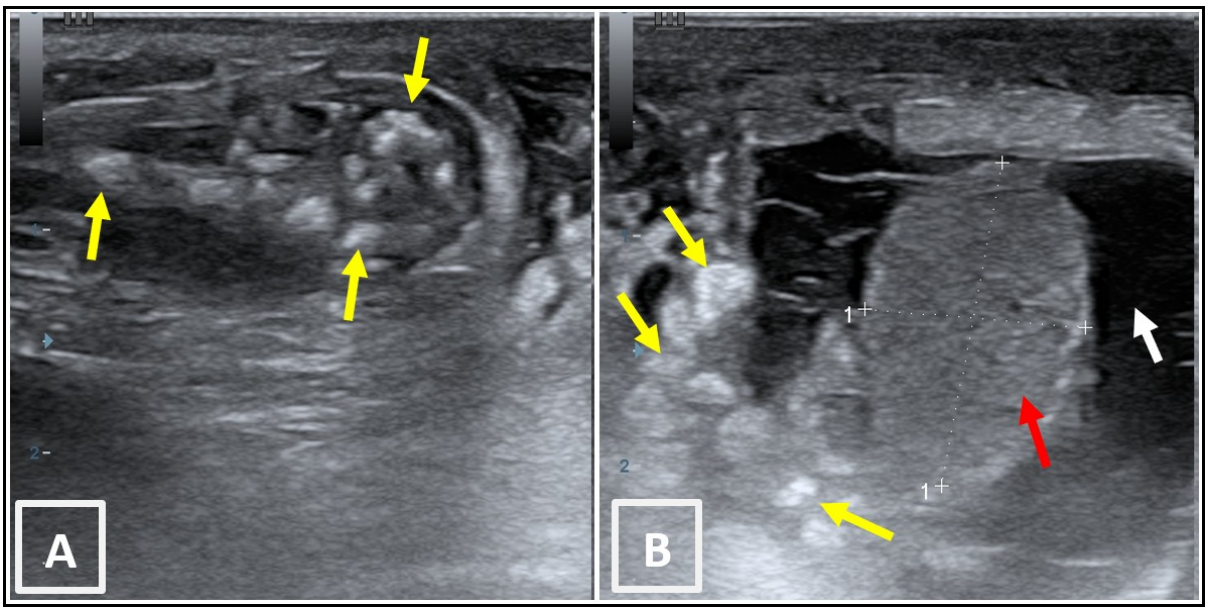

Fig 1. *Scrotal ultrasonographic findings in Patient 1. In the right inguinal region (A), multiple hyper-echoic foci (yellow arrows) were identified. In the left scrotum (B), similar multiple hyper-echoic foci (yellow arrows) were identified, surrounding a normal testis (red arrow) that is surrounded by anechoic hydrocele fluid (white arrow).*

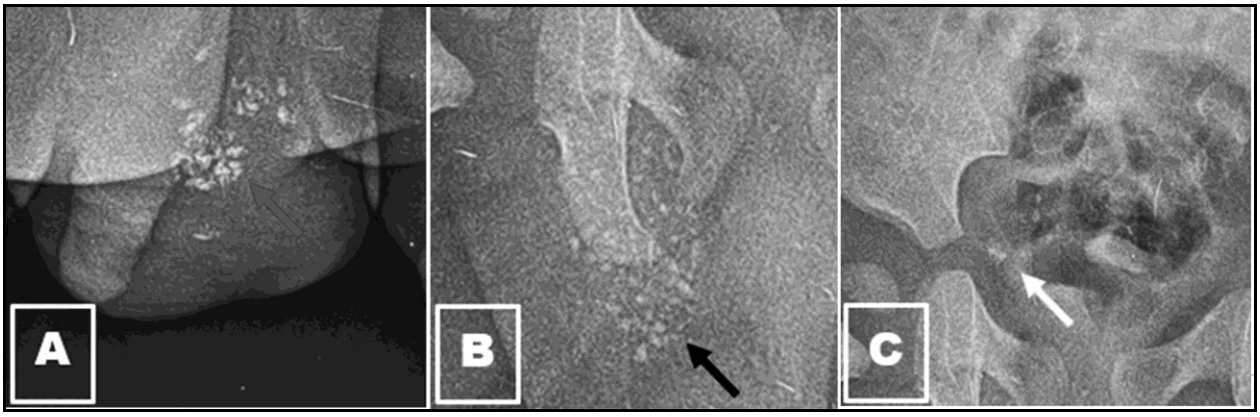

Fig 2. *Plain radiograph of patient 1. Magnified close-up views showing calcifications (arrows) in the left scrotum (A), the right groin (B) and the peritoneal cavity (C).*

CASE REPORT - 1

A 6-month-old infant was referred for acute scrotum. He did not have vomiting, dysuria, fever, or groin swelling. He was born at full-term to a 26-year-old mother. Two prenatal ultrasounds (first and third trimester) did not report any anomaly. At one month of age, parents noticed a left scrotal swelling, which was diagnosed as hydrocele, and the patient was given an appointment at six months for follow-up. There was no history of recurrent respiratory infection or constipation. Before the due date of appointment, he presented with an acute scrotum.

On physical examination, vital parameters were within normal limits. A stony-hard, irregular, and painful swelling of the left scrotum was noted. It was independent of the left testis and there was associated hydrocele. Angell's sign (horizontal lie -

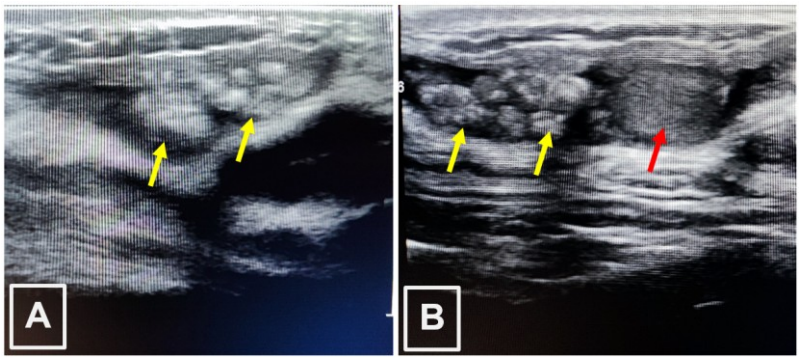

Fig 3. *Scrotal ultrasonography of the patient 2. In (A), hyper-echoic foci (yellow arrows) were identified within the left inguinal canal. However, the processus vaginalis was not patent. In (B), similar foci (yellow arrows) were identified medial to the healthy-looking testis (red arrow).*

of the testis in standing position) was absent. The contralateral scrotum and testis were normal. We suspected acute epididymo-orchitis or MPO. However, testicular torsion and testicular tumor could not be clinically ruled-out. We performed abdomino-pelvic and scrotal ultrasonography (USG) and a plain X-ray. Imaging noted left hydrocele and hyper-echoic (radio-opaque) foci in the left scrotum and in the right inguinal region. (Fig.1) No abnormalities of the epididymis, testes, or spermatic cord were identified.

The plain X-ray identified multiple microcalcifications in the left scrotum, the right inguinal region and the abdomen.(Fig.2) Urine examination did not show any infection. The patient was successfully managed with analgesics upon which pain regressed completely within 3 days. After 12 months of follow-up, he remained asymptomatic; although the left scrotum calcification could still be palpable without any reactive hydrocele.

CASE REPORT - 2

A 5-month-old infant was brought for a small-sized penis. He was born at full-term to a 28-year-old mother. Prenatal ultrasonography had not been done. The mother reported left-side scrotal swelling in neonatal period, which spontaneously regressed a few months later. History of recurrent respiratory infection or constipation was absent.

On physical examination, vital parameters were normal. Penile measurements were within normal range for age according to Park et al.[4] Palpation of the left scrotum revealed multiple, hard, stone-like nodules that were independent of the ipsi-lateral testis. We suspected MPO and performed abdomino-pelvic plain X-ray (Fig.3) and ultra-sonography.(Fig.4) Imaging showed radio-opaque (hyper-echoic) foci within the left scrotum, thus confirming the diagnosis of MPO. No treatment was contemplated for MPO. At two-month follow-up, he remained asymptomatic. He was scheduled for periodic follow-up until adolescence.

DISCUSSION

MPO is a consequence of MP.[5] Due to the patency of the processus vaginalis, natural history of MPO starts with the spilled meconium in the peritoneal cavity that spills into the tunica vaginalis inducing inflammation. Pathogenesis of inflammation in MPO is similar to that of MP.[6] The clinical manifestation of the exudative phase may be neonatal hydrocele. After birth, the natural history of MPO is progressive hardening of the scrotal tissue due to calcification of meconium.[7] As in our patient-1, acute scrotum may be reported during this phase

due to the inflammation resulting from host reaction to the calcifying foreign body.[6] With advancing age, intra-scrotal calcifications tend to regress spontaneously.[7]

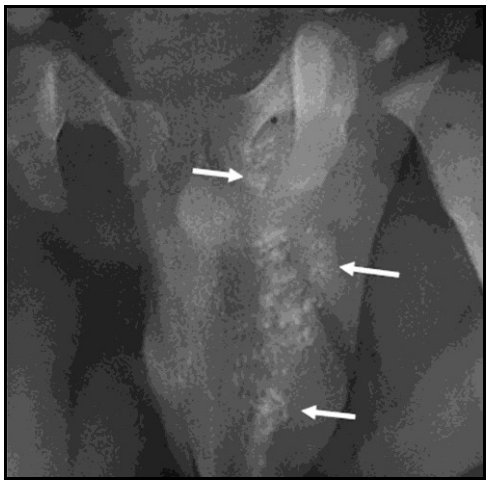

Fig 4. *Abdomino-pelvic X-ray of the Patient 2. Multiple calcifications (arrows) are seen in the scrotum extending into the left inguinal region.*

From 28 weeks of gestation onwards, prenatal diagnosis of MPO was reported in more than 50% of cases using fetal USG or magnetic resonance imaging.[3] However, in resource-poor countries, most pregnancies are unattended or the benefit of regular fetal screening with USG is often absent.[3] Postnatally, the diagnosis of MPO is usually suspected by thoughtful physician as the clinical presentation varies considerably. (Fig.5) It may be an incidental diagnosis either during physical examination or at inguino-scrotal operations for unrelated conditions.[8] When MPO presents as acute scrotum (e.g. acute hydrocele or mimicking testicular torsion), a clinical diagnosis is often difficult. A history of neonatal hydrocele should raise the clinical suspicion of MPO.[3] A history of recurrent respiratory infections may help as it may be linked to the pulmonary manifestation of cystic fibrosis, a common etiology of MP.[6] On physical examination signs of MP should be look for.[6] Scrotal palpation is the cornerstone of diagnosis in isolated MPO with calcification. In our second patient, it guided the diagnostic work-up.

High index of suspicion will guide complementary investigations, which include abdomino-pelvic and scrotal USG or plain X-ray.[9] USG is the imaging of choice due to its specificity and non-invasiveness. The classical diagnostic triad of USG includes hyper-echoic scrotal foci or mass, absence of blood flow through the hyper-echoic mass, and presence of reactive hydrocele. USG also confirms the normal texture of testes.[5,9] However, USG is operator dependant, and in the hands of inexperienced it may be misleading.[3]

In such situations a plain X-ray is useful although radiation exposure of the testis is generally undesirable. Plain radiographs allow easy identification of scrotal and abdominal calcifications. However, visibility of calcified foci depends on the lesion size and the degree of calcification of the lesion.[10] Moreover, USG and X-rays provide insight into the etiology of MP or MPO, especially in neonates.[3]

Management of MPO depends on its nature of presentation. Surgical exploration may be considered when it is associated with other congenital abnormalities (e.g. inguinal hernia, hydrocele, spermatic cord cyst, undescended testis, and scrotoschisis) that warrant an operation on their own merits.[3] Surgical exploration is not indicated in isolated MPO unless a urologic emergency such as testicular torsion cannot be entirely excluded.[3] In a recent review of 18 neonatal cases, we reported conservative management in one-third of patients.[3] However, on retrospective analysis of the indications of surgical exploration in them, we believe that two-third of them should have been conservatively treated.

CONCLUSION

MPO is uncommon; but awareness about it is essential for avoidance of unnecessary scrotal exploration. Its outcome with conservative management is usually good.

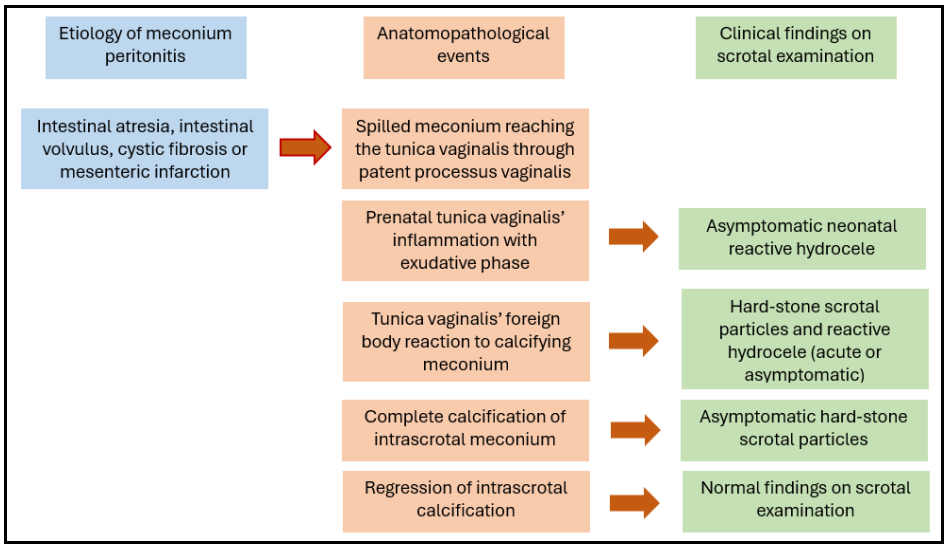

Fig 5. *Natural history of Meconium Periorchitis with varying clinical manifestations*

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Case Report

Giant Para-Urethral Epidermoid Cyst

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Keywords
Epidermoid cyst
Giant urethral cyst
Para-urethral cyst
Scrotal swelling
Urethral obstruction

Abstract

Congenital para-urethral cyst is a rare pathology especially in male infants. We report a large cyst in a 40-days-old infant who was referred suspecting a scrotal tumor. This scrotal mass was present since birth and progressively increased in size to reach 15 x 10 cm. It was associated with dysuria. Attempts at urethral probing were unsuccessful and needle aspiration of the mass yielded a clear liquid. Perineal ultrasound showed a cyst with solid components. The cyst was excised en-block separating its attachment to the prostatic urethra. The postoperative course and short term follow-up were uneventful. Histopathology confirmed it as a benign epidermoid cyst.

INTRODUCTION

Para-urethral cysts are rare in children.[1,2,3] It is commonly reported to arise from the Skene glands of females.[3] In males, it mainly occurs in adolescents and adults, and is secondary to prostatic pathology.[4] The congenital form in infants is uncommon. We report one such newborn with huge para-urethral cyst.

CASE REPORT

A 40-day-old infant was referred to the pediatric emergency department because of a suspected scrotal tumor. The voluminous scrotal mass was present since birth. It was associated with dysuria. It was large at birth and continued to increase in volume aggravating dysuria. His perinatal period was unremarkable.

On admission, he was in good general condition, weighing 5kg. Hyper-pigmentation of the inner

side of the thighs was observed. The scrotal mass of about 15 x 10 cm was found extending onto the penis (Fig.1) and the mass was soft and painless.

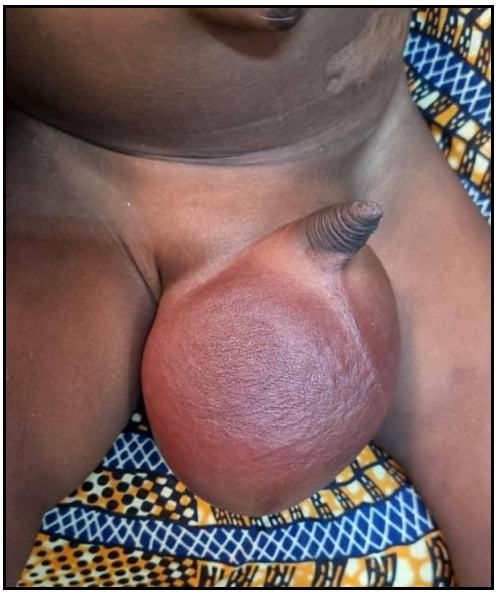

Fig 1. *Voluminous para-urethral cyst*

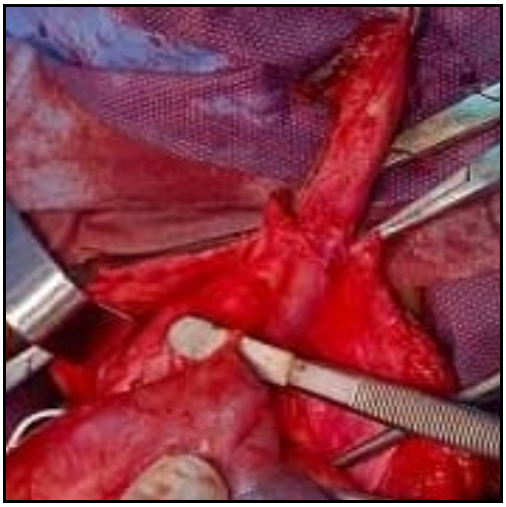

Fig 2. *Pedunculated epidermoid cyst arising from the membranous urethra*

Attempts of urethral probing were unsuccessful and needle aspiration of the mass yielded clear liquid. Cyst or urethral duplication was considered in the differential diagnosis. Ultrasonography revealed a mixed cyst with solid areas. Serum level of beta-human chorionic gonadotropin hormone (0.1µmol/l), serum creatinine (10µmol/l), leukocyte count (10800/mm^3) and hemoglobin (9g/dl) were within normal limits.

Surgery was performed under general anesthesia. By an anterior perineal approach, sparing the skin for the reconstruction of the penis and scrotum, the cyst was mobilized en block to its root. The cyst attached to the membranous urethra by a pedicle was excised. The scrotum and the penis were reconstructed with circumcision. (Fig.3) On follow-up after 2 months of operation, he was healthy. Histopathology was benign epidermoid cyst.

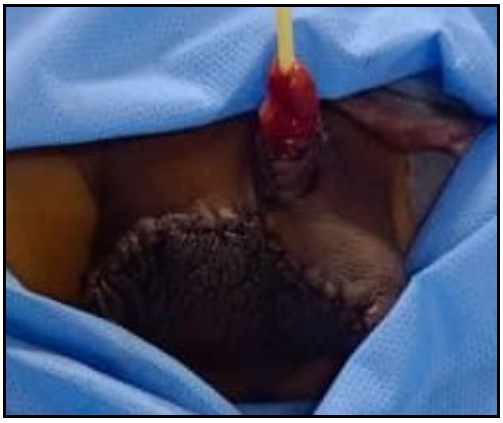

Fig 3. *Image of the reconstructed genitalia*

DISCUSSION

Congenital para-urethral cyst, is a rare condition, especially in boys.[1,2] It may present as a mass lesion or as bladder outlet obstruction. Urethral compression may result in urinary tract infection, urolithiasis, or even renal failure, indicating the potential seriousness of this pathology.[1,2,5] In our patient, the cyst appears to have an atretic communication with the posterior urethra. It was causing partial urethral obstruction although renal function was preserved.

The most common type of para-urethral cyst is meatal cysts. It is seen in adolescence and its presentation is delayed by asymptomatic nature and small size.[2] Our patient falls on the other extreme end of the spectrum of para-urethral cyst.

Our patient presented early because of the anatomical proximity of the cyst to the posterior urethra, its huge size and urethral compression effect. The proximal location (penoscrotal) can be misleading. Because of its huge size, a scrotal tumor was suspected. This was excluded by normal levels of tumor markers and by the cystic nature of the mass. Sub-prostatic location and fibrous pedicle connected with the urethra were also against the features of residual Mullerian cyst.[6] The diagnosis of cystic urethral duplication can be excluded only after histopathology.

Surgical operation of this huge para-urethral cyst is justified by the volume of the cyst and the degree of compression on the urethra that could have long-term repercussion on the upper urinary tract. Operative procedure consisted of complete excision of the cyst along with excision of the overlying excess skin. Treatment options vary according to the clinical presentation and histology, ranging from simple cyst puncture to mutilating surgery.[7] Para-urethral cysts have recurrence rate of about 3%.[7] Complete excision prevents the risk of recurrence.[1,2,7] Epidermoid cyst, although a benign tumor, has a risk of malignant transformation (squamous cell carcinoma) in 2% of patients.[7]

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Case Report

Bladder and Urethral Agenesis: A Report of 2 Cases

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Keywords
Bladder agenesis
Ureterosigmoidostomy
Urethral agenesis
Urogenital anomalies

Abbreviations
CT - Computed tomography
DMSA - Dimercapto succinic acid

Abstract

Bladder and urethral agenesis are among the rarest congenital anomalies, and to our knowledge, only 27 cases have been reported in English literature until now. The authors treated two such cases in the last one year. The first case was diagnosed at 2 months of age, and the second case at 10 months of age. Both were females. In The first case ureterosigmoidostomy was done at 1 year of age. The second case had features of chronic kidney disease due to a stenosed ectopic ureter. Therefore, diverting cutaneous ureterostomy was done. Bladder and urethral agenesis are associated with other congenital anomalies, mostly with upper urinary tract anomalies. To prevent diagnostic delay and to reduce mortality cum mortality by early management, awareness of this rare condition is essential.

BACKGROUND

Bladder and urethral agenesis, one of the rarest urogenital anomalies, has an incidence of 1 in 60,000,000.[1] There are many theories regarding the etiology of bladder agenesis. One of them suggests that it may result from the failure of mesonephric duct and ureteric bud integration in the trigone resulting in lack of bladder filling.[2] Another theory suggested that this may be due to complete atresia of urogenital sinus.[3] In females with absence of bladder, ureter may insert into the Mullerian derivatives such as the uterus, anterior vaginal walls, or vestibule as the Mullerian ducts also open into urogenital sinus.[2] On the contrary, males with the described disease are mostly stillborn, as they can only survive if the ectopic ureters drain into either rectum or patent urachus, which rarely occurs.[4] Surgical intervention to establish adequate urinary drainage is required to protect kidneys from obstructive uropathy and urinary tract infection. Herein, we report two cases of bladder and urethral agenesis in female infants, where both of them had ectopic insertion of the ureter into the vagina.

CASE REPORT - 1

A 2-month-old girl presented with continuous dribbling of urine since birth. A history of periodic normal voiding is absent. Her perinatal period was uneventful. Antenatal ultrasound did not detect any urological anomaly. The anus was in normal position. The external genitalia was that of a female, with a single opening in the vestibule.

Retrograde contrast radiography showed bilateral hydronephrotic kidneys with ureters opening into

a common channel indicating the absence of bladder and urethra.(Fig.1) CT urogram revealed bilateral incomplete duplex pelvi-calyceal system. (Fig.2) DMSA scan showed bilateral renal scarring. Magnetic resonance imaging (MRI) of the lumbar spine did not show any abnormality. The patient was discharged with antibiotic prophylaxis. Subsequently, we did bilateral ureterosigmoidostomy with anti-reflux sub-serosal tunneling. (Fig.3) Double-J stents were kept in both ureters; but both of them got spontaneously expelled within the second post-operative day. The patient was discharged with prophylactic antibiotics and oral sodium bicarbonate as metabolic acidosis was expected. On follow-up after a month, mother complained of incontinent passage of urine per anus, and there were clinical features of metabolic acidosis. However, in the subsequent visits the blood gas analysis showed normal finding.

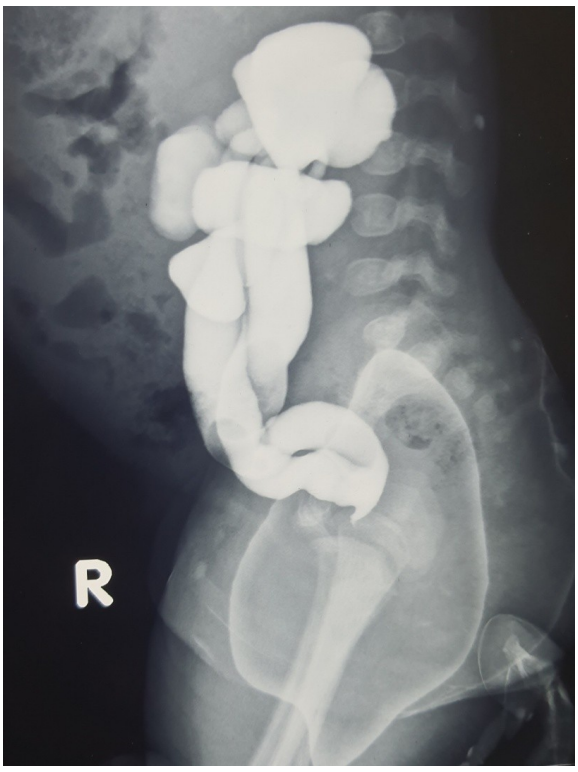

Fig 1. *Retrograde contrast film showing the ureters entering into a common channel in the absence of bladder (Case 1)*

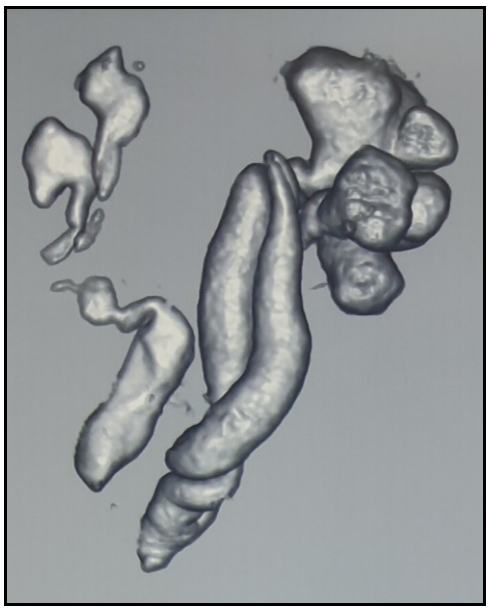

Fig 2. *CT urogram showing incomplete duplex ureter (Case 1)*

CASE REPORT - 2

A 10-month-old girl presented with continuous dribbling of urine since birth without any normal voiding pattern. She had been admitted to hospitals several times for episodes of dehydration, electrolyte imbalance and acute kidney injury which subsequently turned into chronic kidney disease. Prior to the current admission under our care, she had been managed by nephrologists for renal impairment. She had no symptoms pertinent to bowel movements.

During admission, we found her dehydrated and lethargic. Her weight was 5.5 kg which fell below the third percentile. Perineal examination showed a single opening in the vestibule from which urine was coming out. (Fig.4) Spine was normal. Ultrasonography couldn't trace the right kidney and found duplex left kidney with hydroureteronephrosis. In intravenous pyelography, delayed excretion of the left kidney with hydroureteronephrosis and non-visualization of the right kidney were noted. Contrast instillation through the single opening in the vestibule revealed incomplete duplex ureter and pelvis.

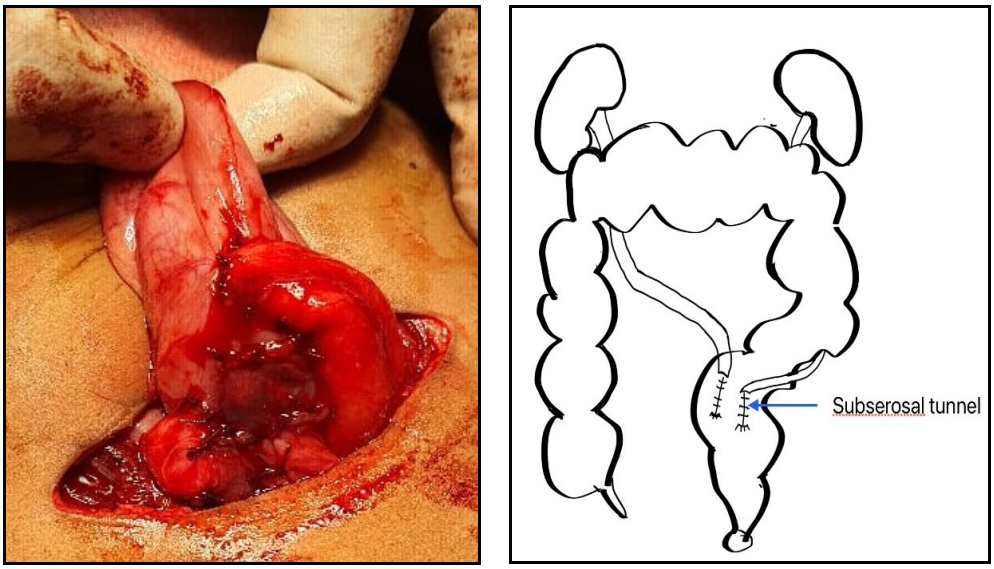

Fig 3. *(A) Intra-operative photograph showing dilated ureters implanted into the sigmoid colon. (B) Schematic diagram showing ureters implanted in sigmoid after creating sub-serosal tunneling*

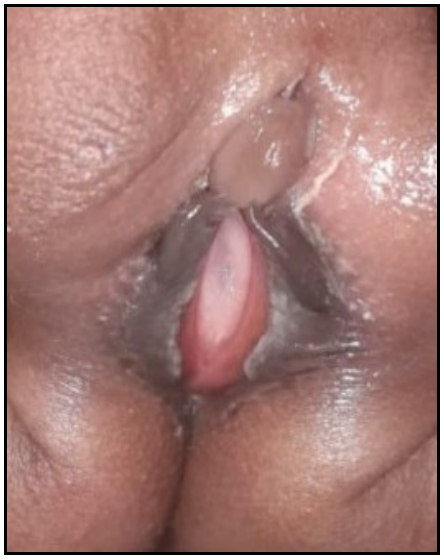

Fig 4. *Single opening in the vestibule (Case 2)*

on the left side. In magnetic resonance urogram, right kidney was not seen in normal position; rather it was located at lower abdominal cavity at level of L3 and L4. The right kidney was smaller and malrotated with anomalous insertion of the right ureter into the neck of small bladder. The left kidney showed multiple small cortical cysts. Left ureter was dilated, tortuous (maximum diameter 2cm) with smooth narrowing at its distal part. Laparotomy revealed absence of bladder and both the ureters opened into anterior vaginal wall. There was a pelvic right kidney with left sided duplex pelvi-calyceal system with grossly dilated ureter. We did urinary diversion by bilateral cutaneous ureterostomy as the patient's condition was not conducive for extensive surgery.

DISCUSSION

Bladder agenesis is thought to occur due to insult at 5-7 weeks of embryogenesis.[2] Most patients with bladder agenesis are females. In males, bladder agenesis is compatible with live birth only if the ureters open into rectum, patent urachus or seminal vesicle.[5] Both of our cases were female and in them the ureters opened into the anterior vaginal wall.

Usually, patients with bladder agenesis presents soon after birth; however, there are reports of delayed diagnosis as late as second and third decades of life.[6] Continuous dribbling of urine and repeated urinary tract infection are the

frequent symptoms. In our patients we initially suspected a commoner pathology, namely ectopic ureter. Nevertheless, absence of normal voiding did not support this initial suspicion. CT Urogram of case-1 was conclusive of bladder agenesis. But, all the investigations of case-2 pointed towards ectopic ureteric rather than bladder agenesis. Seemingly, the dilated vagina was mistaken for small capacity bladder by the radiologists. Examination under anesthesia revealed a single opening in the vestibule. According to Metoki, both of our cases were vaginal subtype of bladder agenesis.[7]

Prognosis of bladder agenesis is variable, and it depends mostly on associated anomalies. Upper tract anomalies such as the solitary kidney, ectopic kidney and unilateral duplex collecting system have been reported. Both of our cases had duplex collecting system. Only one of the previous author mentioned association of bilateral duplex kidney.[8] In addition to this, vascular anomalies and bony deformities has been reported. There are four published cases of bladder agenesis where continent urinary diversion was made, which were either Penn pouch or ileal reservoir along with a Mitrofanoff channel using the appendix. We did ureterosigmoidostomy in case-1 with the expectation that when she achieves bowel control it will also act as a continent diversion.

CONCLUSION

Preservation of renal function, adequate urinary drainage, and continence are aims of the management of bladder and urethral agenesis. Diagnostic dilemmas can delay the initiation of appropriate treatment in these patients. Collaboration of nephrologists, urologist, and radiologists are essential in preventing morbidity and mortality of this rare entity.

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Case Report

Splenic Infarction Due to Acute Malaria in a 10-Year-Old Girl

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Keywords
Falciparum Malaria
Splenomegaly
Splenic infarction
Splenectomy
Complication

Abbreviations
SI - Splenic infarction

Abstract

Non-tender enlargement of the spleen is common in malaria; but splenic infarction is a rare complication. It is frequently masked under acute clinical condition. Until now only a few cases of splenic infarction in patients with falciparum malaria have been published. We describe a 10-year-old girl who presented with signs and symptoms of malaria along with tender massive splenomegaly and worsening pain in the left upper quadrant of the abdomen which was not responding to medications. Contrast enhanced CT scan of abdomen revealed massive splenic infarction. The patient underwent laparotomy and total splenectomy. Her postoperative course was uneventful.

INTRODUCTION

Non-tender splenomegaly is a common finding in malaria. This can lead to complications such as hypersplenism, hyper-reactive malarial syndrome, splenic infarction (SI), and spontaneous rupture of the spleen.[1,2] SI is often masked by the acute febrile illness of malaria.[3] Hyperemia of spleen, plasmodium infection load, endotoxin and allergic response are the pathogenic mechanisms of splenomegaly in acute malaria.[4] The splenic artery is an end-artery with few anastomotic branches. Hence, with increased viscosity of the blood, hemorheology is slowed down, allowing the formation of thrombus and SI. It may be a small segmental infarct or a massive global infarct.[1,5]

Clinically SI patients presents with signs and symptoms of malaria along with severe pain in the left upper quadrant of the abdomen, pain that radiates to the left lower chest and tender splenomegaly.[4] Ultrasonography and computed tomographic (CT) scans can easily diagnose SI.[3] Failure to diagnose and treat it promptly can result in hemorrhage, rupture and abscess formation.[5] Management of SI may be either conservative or splenectomy.[1,6]

CASE REPORT

A 10-year-old girl was admitted with high-grade fever with rigors for 6 days, severe pain, palpable left upper abdominal mass and pallor for 2 days. Fever appeared on alternate days and was associated with sweating and vomiting. Since two days

before admission she developed severe, continuous abdominal pain that was radiating to the left shoulder. Pain was not responding to any medications. Patient belonged to a village of the district that faced outbreak of malaria in recent floods.

On examination she was ill looking and distressed. She was irritable, crying and tossing in agony. Her temperature was 103^0F, pulse rate was 140/min and she was grossly anemic. Lymph nodes were not palpable. The abdomen was scaphoid with a tender firm splenomegaly of 15 x 9 cm. Liver was not palpable. Rest of the physical examination was unremarkable.

Her hemoglobin was 4g/dl, total leukocyte count was 14000/mm^3, platelet count was 48000/mm^3 and erythrocyte morphology was normal. Blood smear was positive for *Plasmodium falciparum*. Ultrasonography confirmed a massive splenomegaly. She was admitted to the hospital and treated with antimalarials, antipyretics, analgesics and blood transfusion. But, her abdominal pain persisted. A contrast enhanced CT scan confirmed massive splenic infarction. (Fig 1)

Patient underwent total splenectomy by open laparotomy. At surgery, the omentum was found to be adherent to the spleen. Large areas of splenic necrosis were noted. (Fig 2) Intra-operative rupture was avoided by careful dissections.

The post-operative course was uneventful and she was discharged on sixth postoperative day after prophylactic vaccination. Histological examination confirmed hemorrhagic infarction of the splenic parenchyma.

DISCUSSION

Pakistan has 1.5 million cases of malaria each year which kills hundreds of thousands of people, especially children under five years of age.[7] Almost all severe cases of malaria are caused by *Plasmodium falciparum* and the spleen is an important host-defense against such infection.[1] Non-tender splenomegaly is frequently seen in malaria and it resolves gradually with standard anti-malarial therapy. Splenic infarction as a complication of malaria is rare. Until now only a few cases of splenic infarction have been reported in young adults with falciparum malaria; but none in pediatric literature. Sub-acute bacterial endocarditis, polycythemia vera, Evans syndrome, sickle cell disease, thrombo-embolism and enteric fever are the other causes of splenic infarction in children.[5,6]

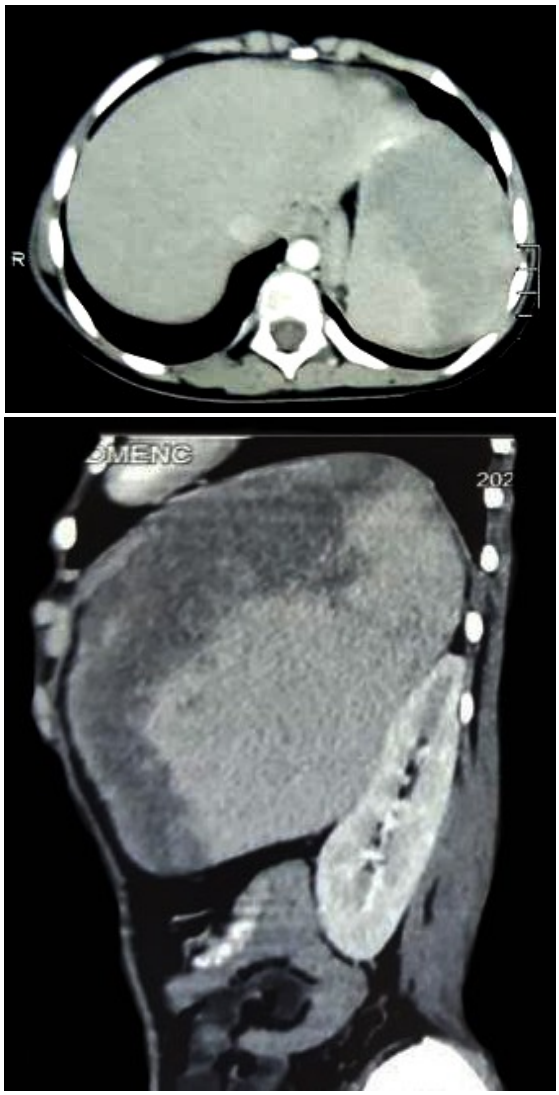

Fig 1. *CT scan showing splenic infarction*

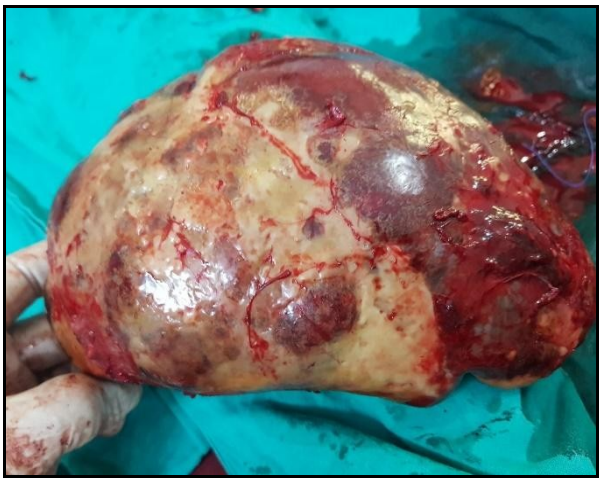

Fig 2. *Resected specimen of spleen showing white infarcted area*

In splenic infarction, the magnitude of splenomegaly remains out of proportion to the lesion. This is attributable to inflammatory edema. The exact mechanism of splenic infarction in malaria is not fully known. However, various theories of pathogenesis have been proposed. They include: (i) hyper-coagulation state, (ii) alteration in the structure of spleen due to adhesion of malaria-infected red blood cells to endothelial cells, (iii) splenic cellular hyperplasia and (iv) anemia-induced hypoxia.[2]

Splenic infarction should be detected during the acute phase of malaria; but it is often masked by the acute febrile illness of malaria.[3] Gupta et.al[4,8] suggested that splenic infarction should be suspected in malarial patients if severe abdominal pain persists in the left upper quadrant, if the pain radiates to the left lower chest or shoulder and if tender splenomegaly occur during the course.

Splenic infarction on ultrasound abdomen is seen as wedge-shaped hypoechoic areas. In our patient ultrasound did not diagnose infarction but was simply suggestive of massive splenomegaly. In diagnosing splenic infarct and its extent, contrast enhanced CT scan has an edge over ultrasound.[6,9]

It is suggested that the antimalarials, analgesics and anticoagulants are effective in treating splenic infarction complicating malaria. But if fever, pain and enlarging tender splenomegaly persist despite adequate anti-malarial treatment, splenectomy is indicated to avoid infarction.[1] Our patient had underwent splenectomy as she was not responding with conservative treatment. Malaria induced splenic infarction shows thrombi in the arterioles, veins and sinusoids associated with hemorrhage, necrosis and infarction.[8] The patient with splenic infarction undergoing emergency splenectomy should receive post-surgical prophylactic vaccination against the *Pneumococci*, *Meningococci* and *Haemophilus influenzae*.

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Tropical Review Series -3

Tropical Phagedenic Ulcer

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Keywords
Fusobacterium
Naga sore
Neglected tropical disease
Non-healing leg ulcer
Phagedenic ulcer
Skin lesion
Synergistic infection
Tropical dermatology
Tropical ulcer

Abbreviations
FU *- Fusobacterium ulcerans*
MMP9 *- Matrix Metallo-proteinase-9*
NTD - Neglected tropical disease
PAF - Platelet-activating factor
TPU - Tropical phagedenic ulcer

Abstract

Tropical ulcer (phagedenic ulcer) is a rapid-onset, slow-to-heal, skin ulcer caused by the synergistic infection of Fusobacterium ulcerans (FU) and spirochetes. It is truly a neglected tropical disease. Confused nomenclature and overlapping clinical features with other tropical skin ulcers led its unpopularity among clinicians and researchers. It is common in warm humid climate. Highest incidence is reported from India and Africa. Ulcers predominantly occur in legs. The source of entry of FU is thought to be minor injuries like thorn pricks and scratches. High amount of cytotoxic butyric acid produced by FU appears to play seminal role in the pathogenesis of ulcers. In acute phase, painful skin ulcers spread rapidly and may involve underlying muscles and tendons. Lesions start as papule which ulcerates in few days. After 1-6 weeks the ulcers become indolent and persist for several months and years. Pseudomembrane covering the floor, putrid magma-like granulation, easy bleeding on handling, foul smelling discharge and extreme pain are characteristic of acute ulcers. Oval or circular shape, cartilage-like indurated base, undermined edges, bund-like raised margins and painlessness are characteristic of chronic ulcers. Diagnosis is mainly clinical because culturing of anaerobic FU is extremely difficult. Leishmaniasis, Buruli ulcer, Yaws, cutaneous diphtheria and ecthyma are the closest differential diagnosis. Chronic ulcers may be complicated by bony erosions and squamous cell carcinoma. Discharge from ulcers has the risk of spreading Hepatitis B. In acute phase systemic administration of penicillins, metronidazole, aminoglycosides and tetracycline along with topical antiseptics are useful. Chronic ulcers require curettage or surgical excision with skin grafting. In modern days death is unusual; but morbidity is considerable. Importance of Tropical Phagedenic Ulcer lies in re-emerging infections from international travel.

INTRODUCTION

Tropical ulcer (phagedenic ulcer) is a rapid-onset, slow-to-heal, skin ulcer caused by the synergistic infection of *Fusobacterium ulcerans* (FU) and spirochetes.[1-3] It is characterized by "an acute-looking ulcer of long duration".[4] Although no age is immune, it is common in older children and younger adults.[1,2] Mortality is extremely rare in

modern days; yet, morbidity is immense and crippling. It is truly a neglected tropical disease (NTD) that the Western scientific world has completely ignored.[5,6] Even the World Health Organization (WHO) failed to recognize it as a distinct entity.[7] A PubMed-based bibliometry of *tropical ulcers* reveals a peak of research activities between 1925 and 1975, followed by a steep decline.(Fig.1) The peak corresponds to the World Wars I and II when the British and American troops returned with these ulcers.[8] The sharp decline in the research output of tropical ulcers may be attributed to its decreasing incidence as a result of better living conditions, good nutrition, easy access to health care, availability of better antimicrobials and modern diagnostic facilities, or it is attributable to a pure apathy of the scientific world. In the last 25 years, there are only fewer than 8 publications on the 'tropical ulcer'![6,8-15] Possibly, many of them might have been mis-labeled as 'simple non-healing ulcers' and treated at non-academic centers without special research consideration. Negligible mortality and morbidity might have also resulted in poor funding chances of research.

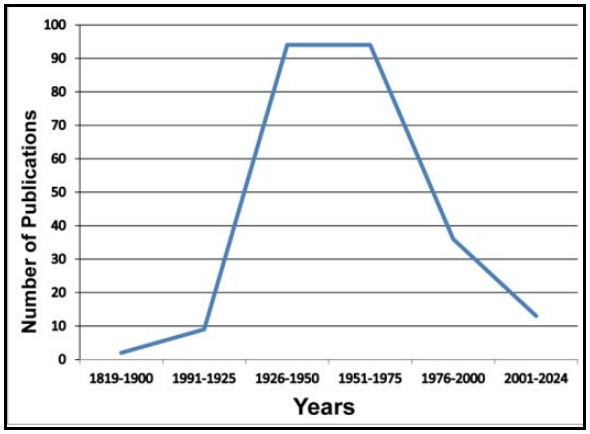

Fig 1: *PubMed-based bibliometry of tropical phagedenic ulcer*

HISTORY

Paleopathological evidences suggest that Copper-age (circa 3000 BCE) people in the basin of the Tanaro River in Northern Italy might have suffered a disease resembling tropical ulcer.[15] *Tropical phagedenic ulcer* (TPU) is well known at least for the last 3 centuries. According to *Jadassohn's Handbook of skin and venereal diseases,* the credit of the first description of *Ulcus Tropicum* in 1792 is attributed to Hunter of Jamaica.[16,17] According to *Harper's Textbook of pediatric dermatology,* Vinson of Mozambique was the first to describe TPU in 1857 among the slaves of merchant ships.[8,18] However, a recent research revealed that the disease was first described by Adriaan Van Brakel in 1774.[19]

Van Brakel was the 'First-Surgeon' of the Dutch East-India vessel *Ouwerkerk* that carried 366 crews to Java. Among them, 63 developed a peculiar leg ulcer which they acquired either while camping at the Cape of Good Hope or while crossing the Indonesian sea and 22% of them died. This unusually high mortality on board prompted the prosecution of Van Brakel for negligence of care. In the era when microbes were not known and antibiotics had not been discovered, what else a naval surgeon could have done, other than dressing the ulcers thrice daily and applying tinctures? In his journal, Van Brakel had accurately recorded the morphology of the ulcers and he noted them as "alkaline, scorbutic and of malignant humour". Thanks to this meticulous description, he was finally acquitted. The juries observed, "Van Brakel's treatment was quite correct, dutiful, and beyond reproach". Interestingly, Van Brakel named them as *Ulcus tropicum phagedaenicum*, (translated into English as Tropical Phagedenic Ulcer) - a term which is still in use. This appears to be the first known detailed description of this pathology. Van Brakel also astutely observed the link between TPU and nutritional by noting that many of those who died were *'scorbutic'*.[19]

Although the pathogenic role of fusiform bacteria and spirochetes had been well known for more than a century, it was in 1987 Beverley Adriaans established TPU as a legitimate disease entity by discovering a new species of bacteria specific to

Box 1: Synonyms of Tropical Ulcer*

Aden ulcer (Corpus 1924)
Annam ulcer (Regnault 1904)
Annamese ulcer (Cras, Laure & Richard 1862)
Annamite wound (Cras, Laure & Richard 1862)
Argentine ulcer (Costa 1944)
Assam Sore (Sen Gupta 1921)
Aurengzebie† (Balfour 1860)
Barcoo rot (Morris & Dore 1913)
Biskra Button† (Paynter 1860)
Bouton d'Aleppe (Murray 1883)
Cachar sore (Sen Gupta 1921)
Chaco ulcer (Costa 1944)
Clou de Biskra† (Murray 1883)
Coast ulcer (Costa 1944)
Cochin sore
Delagoa sore (Apostolides 1922)
Delhi sore† (Balfour 1860)
Desert sore† (Anning 1946)
Drida (Fontoynont 1905)
Doudanduga† (Chinese term)
Epidemic oriental ulcer†
Gallipoli sore
Guadeloupe ulcers (Vincent 1900)
Guyana ulcer (Chapuis 1862)
Jungle sore / rot / ulcer
Kidonda Ndugu§
Malabar ulcer (Corpus 1924)
Malagasy wound (Regnault 1904, Fontoynont 1905)
Mozambique ulcer (Vinson 1856)
Naga (Nagana) sore (Fox 1920)
Natal Sore (Ferguson 1959)
Parangi sore† (Sri Lankan Term)
Phagedenic ulcer (Bartlett 1939)
Phagedenic ulcers of hot climate (Mericourt 1862)
Rhodesian sore (Apostolides 1922)
Shantira (Sen Gupta 1921)
Tokak (Malaysian Term)
Troops (Mooltan) sore† (Murray 1883)
Tropical phagedena
Tropical phagedenic ulcer (Le Dantec 1900)
Tropical septic ulcer (Apostolides 1922)
Tropical sloughing phagedena (Apostolides 1922)
Tropical sloughing ulcer (Cutler 1845)
Tropical ulcer (Wright 1903)
Ulcere annamite (Cras, Laure & Richard 1862)
Ulcere phagedenique des pays chauds (Mericourt & Rochard 1862)
Ulcus tropicum (Hunter 1788)
Ulcus tropicum phagedaenicum (Van Brakel 1774)
Umballa (Ambala) sore
Van Brakel's ulcer (Bruijin 1991)
Veldt sore† (Harland 1901)
Vincent's ulcer† (Stammers 1944)
Yeman Ulcer (Regnault 1904)
Zambesia sore

Compiled from Adriaans,[2] Bruijin,[19] Loudon,[35] Costa,[36] Raynaud [37] and other sources

** Names in parenthesis are either the author who coined the term or those associated with the 'first known' usage of it. Detailed reference list (not included herein) is available from authors on personal request.*

§ In Swahili language it means "my sibling ulcer" as it cannot be easily got rid of.

† Judging from the available descriptions, these terms may be representing a mixed infection of TPU with yaws, Buruli ulcer, Leishmaniasis or cutaneous diphtheria

TPU.[1,2,20-25] He named the new organism as *Fusobacterium ulcerans* (FU).[20,22,25] He was also the first to conduct multicentric studies and electron microscopic studies on TPU.

ETYMOLOGY AND NOMENCLATURE

The term *'tropical ulcer'* has been inconsistently used in the literature with two different connotations.[26] In a *generic sense,* it is used to mean any chronic ulcer seen in the tropics. This includes cutaneous Leishmaniasis, Buruli ulcer, yaws, ecthyma, cutaneous diphtheria and several other venereal diseases.[6,27-30] On the other hand, in a *specific sense,* it is used to mean a peculiar ulcer of the legs caused by the synergistic infection of FU. Still worse is that the term has also been confused with *'trophic ulcers'* (with a misspelling 'tropic ulcers').[31] Therefore, to avoid confusion, the term *'phagedena'* is preferred by some authors.[32] It is a combination of two Latin words, *'Phage'* (to eat) and *'daena'* (avidily) - 'Phagedenic ulcer' means 'a devouring ulcer'. Unfortunately, this term has also been misused to mean any rapidly spreading neoplastic or venereal ulcers.[33,34] Other varieties of hospital-acquired necrotizing fasciitis such as Fournier's gangrene, Cullen's ulcer and Meleney's

ulcers have also been referred to as phagedena.[35] Adding to the confusion, this pathology is also known by a variety of geographic and local names that are shared with other pathologies.[1-3,19,36,37] (Box 1) For example the term *Delhi sore* is applied to both TPU and cutaneous Leishmaniasis.

Interestingly, in pre-independent India, TPU was also known as *Aurengzebie,* with reference to the Mogul emperor *Bahadur Alamgir Aurangzeb* (1618–1707 CE).[38] The etymology of this word is ascribed to the sufferer, which is variously cited as the emperor himself, the British army stationed at Delhi cantonment and the citizens of Delhi.[39] It may also be a contemptuous expression maligning the tyrannical emperor. On retrospective analysis, *Aurengzebie* appears to be a mixed lesion of TPU and cutaneous Leishmaniasis.

From the foregoing it is evident that one has to be extremely wary of the confused terminologies used in the published literature.[3] Imprecise terminology and overlapping clinical features of TPU with several other pathologies prompted Clement to comment that *tropical ulcers* were the "diagnostic garbage heaps of tropical medicine".[3] The descriptive term *tropical phagedenic ulcer* (TPU) coined by Le Dantec in 1900 appears to be less confusing and hence is preferable.[26,40,41]

ETIOPATHOGENESIS

Several etiopathogenic theories of TPU have been proposed. Many of them are anecdotal without proper research evidence. It is now established that TPU is essentially an infectious disease. What predisposes to the infection is extensively debated in the literature. Marsh and Wilson succinctly summarized the etiopathogenesis of TPU as "Filth (Flies), Food, Friction and Fusospirillosis".[42]

Nutritional Deficiency Theory

Dietary deficiency is the oldest pathogenic theory of TPU known since 1774. Van Brakel considered it to be a consequence of vitamin C deficiency.[19] During the Child's War (1686–1690), the British troops sieging Delhi did not develop TPU thanks to the availability of good food, while inhabitants of the inner city including the emperor Aurangzeb were said to have developed the disease owing to a short supply of nutritious food.[30,39] Approximately 30% of TPU patients are malnourished.[43] McCulloch called TPU a '*dietetic ulcer'* because adding fish to the diet reduced its incidence.[44] Deficiencies of vitamin B-complex[48,49] (especially riboflavin), calcium[3,44-46], zinc[47], vitamin A,[48] and animal protein[44] have been implicated in the pathogenesis of TPU.

Nutritional deficiency theory is supported by the high incidence of TPU during drought seasons and during rainy seasons when fishing activities are abandoned. The rich people in cities are rarely affected as compared to the rural poor. However, several authors disputed and disproved this theory.[2,41,42,47,50-53] Interestingly, the ulcers healed faster in patients treated with topical cod-liver oil dressings than in those who were given the fish oil orally.[48] Irrespective of its role in primary TPU, nutritional deficiencies certainly increase the risk of recurrent ulcers.[48]

Vector-born infection theory

James observed that TPU is very common in areas endemic to malaria.[54] He proposed that TPU could be a vector-born disease.[48,55] Nevertheless, Loewenthal observed that malaria was common in both rich and poor, while TPU was exclusively seen in the poor.[44] Abrasions caused by scratching of insect-bite sites probably serve as the entry point of the infective agents.[55] Mosquitoes, flies and leeches may act as mechanical vectors of pathogenic microbes. [56,57]

Traumatic Theory

It is generally accepted that tiny abrasions and minor injuries facilitate the entry of pathogens. The injury may be trivial enough to be easily forgotten by the patients. They may be thorn

pricks, scratches, insect bites, scabies or sports injuries. Nearly 24-94% of patients had a history of injury.[2,52] Apostolides reported a nurse who developed TPU of finger after accidental pricking with a contaminated scalpel.[58] The traumatic theory is supported by the fact that TPU is common in the legs of boys who play outdoor games without wearing protective full trousers. The high incidence of TPU in agricultural farms and in barefoot walkers is also linked to trivial injuries. Interestingly, the sole of the foot is never involved even in barefoot walkers, perhaps due to thick epidermis.

Vascular Insufficiency Theory

The high frequency of TPU in the lower leg raised a suspicion if it could be a form of vascular insufficiency.[3,59] Jackson suggested that the ulcers may be caused by interrupted dermal blood supply as a result of obstructed perforator vessels as they pierce through the deep fascia.[2,60] Findings that support the vascular theory are presence of intravascular thrombosis near the ulcer, thickening of the vessel wall by endothelial hyperplasia, perivascular infiltration of inflammatory cells, septic emboli within the vessels and degeneration of vessel walls in the granulation tissue. However, Adriaans found no evidence of vasculitis in histopathology and electron microscopy.[2,21] He attributed the vascular changes to the effect of healing and inflammation.

Synergistic Infection Theory

Bacterial synergy is the widely accepted theory of TPU pathogenesis.[24,25] It was first proposed in 1899 by Le Dantec, Plaut and Vincent.[2] Although the association of fusiform bacteria with TPU was well known for several decades, in 1986 Adriaans discovered that the isolated species is unique to this pathology and named it as *Fusobacterium ulcerans*.[20,22,23,25] In addition to FU, *Bacteroides*, coliforms or *Treponema vincentii* (a spirochete) are also frequently isolated from the ulcers. Pure isolates of these organisms did not cause ulcers when injected into guinea pigs or rabbits;[2,53] but when injected together, they caused typical TPU in animals and human volunteers, thus satisfying the Koch's postulates of infectivity.[1,2,53] These experiments indicate that these organisms are less virulent to cause disease by themselves and when inoculated together act synergistically. The low virulence and host immunity could be the reason for the spontaneous halting of ulcer size after the initial phagedenic phase. The presence of FU as a commensal in the oral cavity and intestine in 25% of TPU patients has lead to the hypothesis that they may be spreading by auto-inoculation from eating with the hand, by public spitting of saliva, or by the topical application of native medications prepared by mixing saliva or cow dung.[3,58]

Cytotoxic Enzyme Theory

Although FU is isolated from TPU, the exact pathogenic mechanism is not known. A hypothesis of bacterial metabolic by-products causing cellular damage has been proposed.[60] FU produces a high amount of butyric acid which is proven to be directly cytotoxic to Vero cell lines *in vitro* at a concentration as low as 0.005M.[23] Heparinase synthesized by *Bacteroides* causes intravascular clotting thereby interrupting blood supply to the ulcer area.[60] The resultant anaerobic environment favors the growth of FU, thus establishing a synergy.

Apart from the bacterial exotoxins and endotoxins, dying host cells also liberate several autodigestive enzymes such as extracellular protease, collagenase, elastase, lecithinase, hemolysin and lipo-polysaccharides. These enzymes may play a major role in the pathogenesis of TPU. Platelet-activating factor (PAF) when combined with bacterial lipo-polysaccharide can cause extensive tissue necrosis.[61] Biosynthesis of PAF by host immune cells is stimulated by bacterial endotoxins and hypoxia. PAF recruits more cellular elements which in turn synthesize more PAF. As surgical excision of the ulcer removes the source of toxic

enzymes, TPU are found to heal faster with this treatment than with antimicrobial therapy.[60]

Recently, proteolytic enzymes such as matrix metalloproteinase-9 (MMP-9) released by FU are shown to inhibit wound healing. They cause destruction of extracellular matrix and inactivation of epithelial growth factors.[62] Interestingly, MMP-9 is a calcium-dependant and zinc-containing endopeptidase, thus reviving the interest in nutritional theory of TPU.

Comorbidity Theory

TPU is known to occur as a superadded lesion to an already existing leg ulcer such as that of sickle cell disease, Leishmaniasis, Buruli ulcer or yaws. The frequent association of TPU and hepatitis in Kiribati and Gambia suggests that both the diseases could have been acquired through a common mechanism, namely trauma.[63,64] As FU is a commensal in oral cavity, gingivitis is proposed to be a risk factor for TPU.[43] James believed that Malaria is a predisposing factor of TPU.[54] The postulated pathogenic mechanisms include hemolysis liberating iron needed for bacterial growth, immunosuppressive effect of splenomegaly, toxins released by plasmodium, mosquitoes acting as a mechanical vector, protein catabolism of malarial fever reducing the host immunity and the introduction of infection by scratching the mosquito-bite site[54] However, Adriaans did not find any significant pathogenic association of TPU with comorbidities.[2]

Genetic Theory

Coloured races (Africans and Indians) and Chinese are more commonly affected by TPU than the White races. It is more common among the Nuba tribe of Sudan than in Arabs.[48] Among Kenyans, Kikuyuns are more prone to TPU than the Maasai tribes. Similarly, Rwandans are more affected than the Baganda tribes of Uganda.[48] In Assam, it is common in the Uriah caste.[57] These differences may not be due to genetic susceptibility; but rather to cultural variations in dietary habits, exposure to causative organisms and lack of protective footwear. This is attested by the fact that the British and American troops, deputed to tropical terrains during the two World Wars, developed TPU.

DEMOGRAPHY

Geographic Distribution

TPU is mainly reported from the tropics and subtropics.[1] It is endemic in Gambia,[47] Zambia, Central Africa,[2] Uganda,[65] Zimbabwe,[66] South Africa,[18] Mozambique, Nigeria,[67,68] Ethiopia,[69] Kenya, Sudan, Libya, Southern India,[70] Eastern India,[56,57] South China, Indonesian archipelago, Vietnam, Caribbean islands,[71] Fiji,[14] Australia,[3] Gold coast, Papua New Guinea,[72] Bolivia, Brazil and Argentina.[2] (Fig 2) It is more common in the rural areas of Gambia than in the urban centers.[44] Well-documented outbreaks[12,73-75] (Box 2) and travel-acquired TPU have been reported from other parts of the world including temperate countries.[6,14,36] Although TPU is nicknamed as *desert sore*, it is rare in the Arabian Peninsula. It appears that a hot humid climate rather than a dry torrid environment is essential for the pathogenesis of TPU. For this reason, it is common in the cold highlands of Somalia and in Assam with the highest rainfall in India.[1,2]

Incidence and Prevalence

The exact incidence of TPU is not known. Several of these patients might have been treated by native healers without a diagnostic label of TPU. In Madurai (Tamilnadu), 10 new cases were seen daily at a smaller clinic located on the banks of the Vaigai River, while there were none at a nearby big teaching hospital that had a footfall of 3,000 new patients per day.[41] This illustrates the case selection attitude of hospitals in the same geographic location. TPU was endemic in the tea estates of Assam, they were never seen at the Tata Tea Estate of Munnar, although the work environment

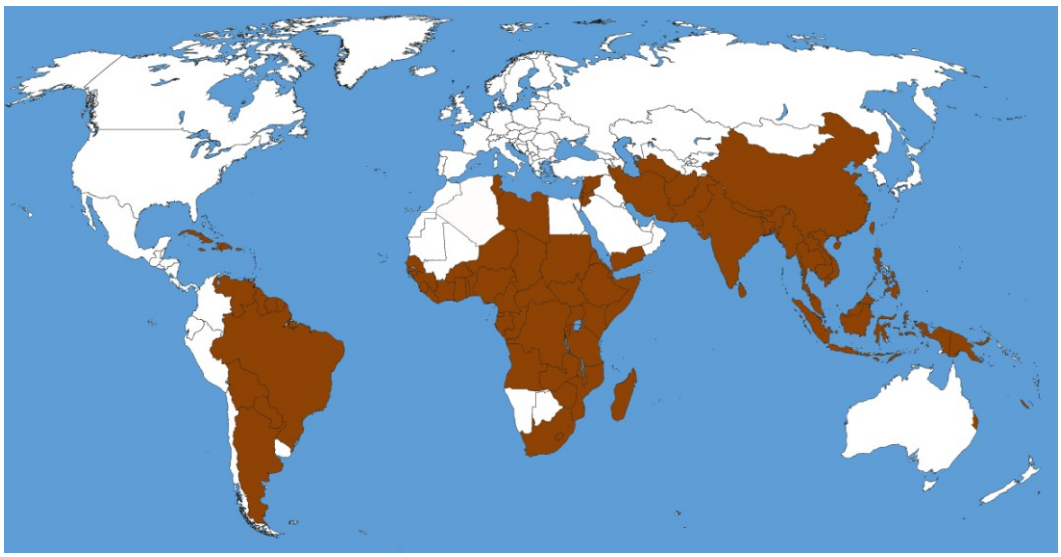

Fig 2: *World-wide distribution of Tropical Phagedenic Ulcer (Compiled by analyzing 100 years of literature, 1924 - 2024)*

Box 2. Recorded Outbreaks of Tropical Phagedenic Ulcers

Geography	Year	Reference
Assam, India	1908	Patterson 1908
Transvaal, S. Africa	1911	Lister 1911
Palestine	1919	Apostolides 1922
Assam, India	1920	Fox 1920
Unnao (UP), India	1920	Mathur 1922
Syria	1923	Adams 1923
New Hanover Island	1936	Clements 1936
Solomon Island	1938	James 1938
Algeria	1943	Bertrand 1950
Calcutta, India	1943	Panja 1943
Natal, South Africa	1958	Ferguson 1959
Cooks Islands	1976	Kuberski 1980
Chennai, India	1977	Yesudian 1979
Djibouti	1997	Kerleguer 2003

of both places were identical.[41] Tumwine from Zimbabwe reported the largest series of 1,680 patients collected over 14 months.[66] Among the 36,000 children admitted to a Nigerian hospital over 10 years, there were 126 cases (0.35%) of TPU.[76] Prevalence at a refugee camp was 6.9% and at a primary school was 8.2%.[41] In a community survey, TPU forms 7.4% of all skin infections in Ethiopia[66] and 0.4% of all chronic leg ulcers in the tropics.[77] About 3-7% of hospital admissions in Sudan[48] and about 95% of skin ulcers in Nigeria are due to TPU.[1] It is found to occur in 1% of the vulnerable population in Uganda.[65] Very high incidence to the tune of 33-50% of hospital out-patients has been reported from Nigeria and Uganda.[44] There are some indications that the annual incidence of TPU is progressively falling in the last 4 decades.[2]

Age Distribution

Although TPU can occur at any age, it is common between 5 and 15 years (mean 9 years).[1,2] It is extremely rare below 5 years and above 45 years of age.[1,2,41,78] As young infants are carried by mothers, they are unlikely to contract infection from environmental injuries. The youngest patient reported was a 3-years old infant.[79] The reason for the high prevalence in older boys is attributed to outdoor gaming activities without protective full-length trousers or footwear. James reported 2% of TPU occurring in infants below 2 years of age.[54] In India 41% occurred in age less than 15

years, 35% in 16-30 years, 21% in 31-45 years and 2% in above 45 years of age.[57]

Sex Distribution

The sex distribution of TPU is variously reported as equal ratio,[1,2] a female preponderance,[44] or a male predominance. A moderately high male ratio of 2:l is reported from India and Trinidad;[45,51] while others [52] found a male predominance in the cities and an equal sex ratio in the rural areas. A very high male ratio of 14:1 has been reported from India.[2,41] The 93% male frequency reported by Buchanan is exceptional.[80] The variations in sex incidence are attributed to the nature of occupation exposure to injuries. For example, men who do fishing from boats are less prone to TPU than women who do agricultural activities in marshlands.[2]

Seasonal Variations

In Aden, Sumatra, Gambia, Zambia and West Indies, TPU is common in the wet rainy seasons. However in Libya, it is common in dry hot draught seasons.[2] Seasonal scarcity of certain foodstuffs and nutrients could be the reason behind this variations. It is suggested that TPU is common in warm humid periods of the year due to excessive sweating, skin maceration and itching-scratching injuries that leads to inoculation of pathogens.[52] Fresh lesions erupt after a brisk shower followed by bright sunshine.[2] Therefore, a warm humid atmosphere appears to be essential for the proliferation of causative organisms in the environment. Few authors from India and Papua New Guinea have denied any seasonal variations in the incidence of TPU. [45,50]

Contagiousness

Loewenthal [44] and Adriaans[2] have proved that person-to-person spread is rare. Epidemics of TPU appear to have been due to direct inoculation from environmental rather than from human contacts. Overcrowding is not a risk factor, as TPU does not occur in several members of the same family.[44]

PATHOLOGY

The histopathological features of TPU are non-specific.[2] They vary according to the stage of the disease, the effect of treatments and the zone of ulcer that is biopsied.

At the early stages, a coagulum containing necrotic tissues, exudates and bacteria forms a pseudomembrane resembling that of diphtheria. Underneath the pseudomembrane, bacteria are arranged in a palisade fashion. Histopathological features of the acute phase include spongiosis (intercellular edema of the epidermis), elongation of the rete ridges, polymorphonuclear leukocytic infiltration, destruction of melanocytes, edema of the dermis, disorganized collagen strands, dilated capillaries, hemorrhagic foci and micro-abscesses at the adjacent soft tissues. Acanthosis (thickening of the stratum spinosum) at the peripheries of the TPU causes raised pigmented edges.[1,2]

In the chronic phase, progressive fibrosis of the ulcer base, peripheral hyperkeratosis (thickening of the stratum corneum), pseudoepitheliomatous hyperplasia, foreign-body giant cells reaction and lymphocytic infiltration are seen. Arteries show marked hyperplasia of smooth muscles and endothelium resulting in narrowing of lumen.[2] Some authors consider these vascular changes as the primary pathology rather than the effect of inflammation.[2] Adriaans has conclusive excluded vasculitis in the pathology of TPU.[2] Nerves are not affected.

Electron microscopy (EM) confirmed the features of conventional histology.[21,81] Direct bacterial cell lysis seen in EM may explain the rapid necrosis of tissues in the acute phase. EM demonstrated an equal proportion of helper and suppressor T cell infiltrations, but only a few B cells.[21]

Box 3. Organisms Isolated From Tropical Phagedenic Ulcer

Coliforms (60%)
Fusobacterium ulcerans [1] (35-40%)
Lysinibacillus fusiformis [2] (30%)
Bifidobacterium (30%)
Staphylococcus epidermidis (30%)
Peptococcus (23%)
Peptostreptococcus (23%)
Bacteroides (20%)
Streptococcus § (15%)
Enterobacter cloacae (14%)
Enterococcus faecalis [3] (12%)
Citrobacter freundii (10%)
Pseudomonas (7%)
Propionibacterium (8%)
Treponema vincentii [4] (5%)
Proteus sp (3%)
Escherichia coli (2%)
Staphylococcus aureus (1.6%)
Veillonella (0.8%)
Corynebacterium ‡
Haemophilus ducreyi
Yeast (Candida)

Data Source: Adriaans[2,24,25]
Previous nomenclature:
[1,2]Bacillus fusiformis, [1]Leptotrichia buccalis, [3]Streptococcus fecalis, [4]Spirochaeta schaudinni, [4]Borrelia vincentii

§ Includes hemolytic and non-hemolytic subtypes.

‡ This appears to have been isolated from a case of mistaken clinical diagnosis. However, TPU is known occur as superadded infection to already existing cutaneous diphtheria or Leishmaniasis.

MICROBIOLOGY

The microbiology of TPU is faced with several challenges: (i) Anaerobic culture facilities to grow FU are not universally available. (ii) Culturing of *Treponema* is extremely difficult. (iii) Dark field microscopy which is essential to visualize spirochetes is not available in many low-middle income centers. (iii) Secondary invaders and surface contaminants often mask the original pathogens. Costa asserted that contaminants are usually superficial while actual pathogens are located in deeper layers.[36] However, this is not scientifically proven. (iv) Isolates differ according to the site of sampling and pathogenic phase.(Box 3) Contaminants are more at the center of the ulcer while true pathogens are at the advancing peripheries. Cultures were positive in 36% within 6 weeks of onset, while only in 1% after that.[2] (v) Several anaerobic pathogens die during the transport of samples thus accounting for a high percentage of negative cultures.

The primary pathogen of TPU is FU. They are non-sporing pleomorphic Gram-negative obligate anaerobic rods that produce large amounts of butyric acid. Among the 15 varieties of Fusobacteria, different species are etiologically associated with different diseases.[82] For example, *noma* is caused by *F.necrophorum*, gastric ulcer by *F. gastrosuis* and ulcerative colitis by *F. nucleatum* and *F. varium*. Similarly, *F. ulcerans* [20,22] and *F. nucleatum* [81] are identified to be specific of TPU.

Two distinct morphological variants of FU have been described. Group-1 FU (NCTC 12111) is coccoid, forming huge dome-shaped creamy or yellowish smooth colonies. They have pointed ends and are capable of growing in selective medium with bile. Group-2 FU (NCTC 12112) is irregularly stained, long rod-shaped or filamentous, forming tiny flat colonies. They have bulbous swelling in the center of the rod.[20] Both variants are commensals in the oral cavity and colon.[83] The Group-1 FU resembles *F. varium,* while the Group-2 FU resembles *F. moriferum*. However, they differ in their fermenting characters.

Pre-reduced anaerobically sterilized, peptone yeast extract (PRAS-PYG) agar and broth dispensed into Hungate tubes are the ideal transport medium for FU.[25] It allows successful culturing as late as 6 weeks of sampling.[2,25] Eh indicator in the medium will reveal any break in the anaerobic

seal. Brain heart infusion (BHI) broth or agar is the recommended regular culture medium for FU.

Pathogenic source of FU is variously described as saliva, mud, thorny bushes and marshland.[41] A minimum inoculum size of 10^{12} Fusobacteria was needed to cause the lesion.[2,20,24] FU is never seen in histological sections probably because of small numbers.[50]

CLASSIFICATION

TPU may be primary (de novo) or secondary. It may occur as a superadded infection of guinea worms, scabies, cutaneous Leishmaniasis or dog bites.[45] Secondary TPU has often led to much confusion in the literature. From epidemiological point of view, TPU may be classified as sporadic, endemic or epidemic.

STAGING

Based on the evolutionary characteristics of the ulcers, TPU is variously staged by several authors. Innes[4] classified them into acute, sub-acute and chronic types; Pattanaik[84] into early, sloughing, indolent and healing stages; Blaine[5] into acute, chronic-on-acute and chronic indolent ulcers; and Robinson into phases 1 (pre-ulcerative papule), 2a (acute ulcer), 2b (chronic ulcer) and 3 (healed ulcer).[2,41] In 1938, James[54] classified TPU into Stage 1 (active progressive ulcers), Stage 2 (sub-acute ulcers with cessation of progression), Stage 3 (clean, chronic or healing ulcers). In the same paper, he also provided another detailed staging system which is modified and adopted in this review. (Table 1)

CLINICAL FEATURES

Clinical features of TPU vary according to the stage of the disease.

Stage 1 (Prodromal or Pre-Ulcerative Stage)

Pre-ulcer stage lasts for a few days. A small papule of a few millimeters size develops at the site of inoculation and it soon becomes a vesicle and pustule.[78] It is highly painful and may ooze sero-sanguinous fluid. Only 3% of patients present at this stage.[41,78] These lesions are usually seen by physicians in the form of a satellite lesion that erupts near the existing ulcers. Within a few days black discolouration occurs around the papule. Rarely, the vesicle may transform into a hemorrhagic bulla. At the end of one week, the central area necroses, leaving behind a tiny ulcer.

Approximately 80% of the lesions occur between the knee and the ankle.[1,2,57,78] The dorsum of the foot, thighs and toes are also frequently involved. The sole of the foot is never affected.[57] (Fig 3) Rarely, it may occur in shoulders, arms, trunk, neck, and scalp.[2,50]

Table 1. Staging of Tropical Phagedenic Ulcer

Stage	James Staging (1938)	Modified Staging (2024)
1	Idiopathic ulcer †	Prodromal stage
2	Tiny ulcer *	Acute phagedena
3	Phagedenic ulcer	Chronic indolent ulcer
4	Chronic ulcer #	Chronic erosive ulcer
5	Recurrent ulcer ‡	Healing (Cicatrix)
6	-	Recurrence §

*† It means the antecedent minor trauma; * Caused by ruptured vesicle; ‡Includes even remote ulcers, reactivation of infection; §Includes only recurrent ulcer at the healed site. Ectopic recurrences are considered as separate episodes of reinfection. Reactivation of healing ulcer is considered a regression of stage; # James termed it as "Cessation of phagedena"*

Stage 2 (Acute Phagedena)

It lasts between 1 to 6 weeks of onset. About 65% of TPU patients are seen within a month of onset.[41] At this stage, the ulcer rapidly spreads in all directions. It is highly painful and is covered by a thick tenacious, gray-green or ash-white, fetid slough resembling that of diphtheric pseudo-membrane. Copious discharge attracts flies. The floor of the ulcer is filled with crimson-red, friable granulation tissue that bleeds easily on touch. It is graphically described as putrid magma-like.[36]

(Fig 4) The ulcers are oval, circular or of a regular geometric pattern with undermined edges, raised borders and black discolouration of peripheries. There may be mild edema around the ulcer; however adjacent tissues are mostly unaffected.

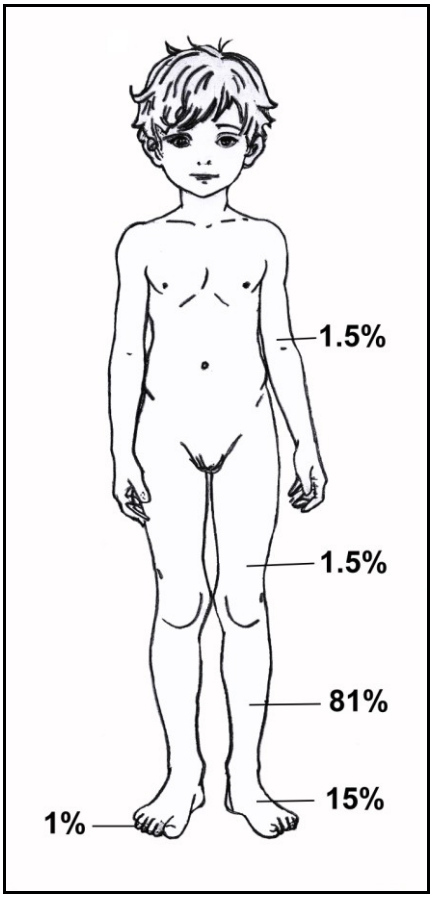

Fig 3. *Anatomical distribution of Tropical Phagedenic Ulcers (Drawn using Pikaso AI tool; retouched by Srinidhi) Data source: Adriaans* [2]

The ulcers are usually single (75-80%)[52] or occasionally multiple (25%). [1,2] As many as 8 ulcers have been reported in a single patient.[57] Kissing and satellite ulcers may coalesce into a single large ulcer. The size of the ulcer is usually 2 to 6 cm; but ulcers as large as 15 cm in diameter have also been reported in 6% of patients.[1,2,41] Exceptionally, Kolawole reported an ulcer of 40 cm diameter that encircled the limb.[67] The depth of the ulcer is usually less than 2 cm[41] and is inversely proportional to the extent of ulcer.[85] During the phagedenic phase, the infection may involve the underlying muscles, tendons and bone. The ulcer heal slowly over 2-6 months.[44]

Systemic symptoms are either absent or minimal. However, during the first two stages, mild fever and malaise occur especially in young children. Lymphangitis, erysipelas and regional lymphadenopathy are seen in 25% of TPU.[5,86] Acute lymphangitis may cause massive edema of the lower limb resembling elephantiasis.[5]

Stage 3 (Chronic Indolent Ulcer)

This stage lasts from 6 weeks to several months (range 2-6 months). Rarely, they defy healing and persist for several years. MacDonald cited an unusual case of ulcer that persisted for 18 years![6] A peculiar mucoid secretion gives a glazed appearance to the ulcer. Ulcers at this stage are stabilized in size and are painless; but rarely, nocturnal pain may be experienced.[36] Secretions are minimal and non-foul smelling. The base of the ulcer is indurated and hard that it resembles a cartilage on palpation. A gritty sensation of the base is appreciated when a needle is pierced through it while injecting local anesthesia for biopsy. The edges are more prominent, sclerotic and elevated resembling a bund. This soft tissue thickening is the hallmark of TPU. The floor is relatively clean with a pale-pink granulation which is less friable and does not bleed readily on palpation. Rarely, the chronic ulcers exhibit periodic exacerbation with further expansion of the ulcer size.[60] Jackson believed that TPU do not involve deep fascia and such involvements are almost always due to superadded infections.[60] With secondary infection adjacent tissue may become erythematous for a considerable distance.

Stage 4 (Chronic Erosive Ulcer)

TPU is predominantly a disease of the skin and soft tissues.[44] However, it may also erode into the underlying tendons, muscles and bone.(*Vide infra*) This stage may last from 6 months to 10 years.[5]

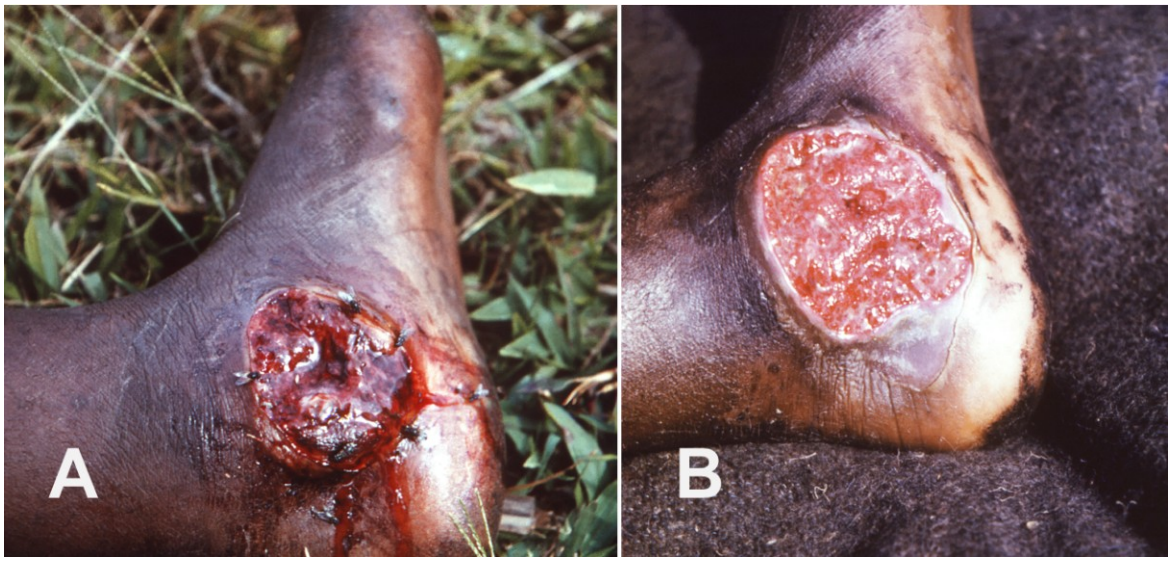

Fig 4. *Clinical appearance of Tropical Phagedenic Ulcer. (A) Acute phagedenic stage; (B) Chronic non-healing stage of the same patient. (Public domain figures from Public Health Image Library ID No. 12174 and 12178 respectively)*

Stage 5 (Healing & Cicatrization)

At this stage, healing starts by flattening of the edges. The resultant scar resembles the shape of the original ulcer. The scar is thin and pale-white resembling a parchment paper. (Fig. 5) The mean duration of healing is 20 weeks (range 1-52 weeks). However, indolent ulcers may persist for 12-15 years.[5,8] As the scars are atrophic, they are liable for repeated breakdown and recurrent ulcers. The causative organism is said to lurk in the scar with periodic reactivation.

DIAGNOSIS

Diagnosis of TPU is mainly based on the clinical characteristics of the ulcer. Flora Innes[4] defined 3 diagnostic criteria as follows:

1. Acute-looking ulcer of long duration. (Punched out edges, undermined margins, glary mucoid foul smelling discharge, free bleeding and pearly islets projecting through the base)
2. Extreme pain in the acute and sub-acute stages
3. Demonstration of *Fusobacterium* with Spirochetae in the ulcer

Dark field microscopy, anaerobic culturing and radiographs of underlying bones are helpful in the diagnosis. Neutrophilic leukocytosis, elevated C-reactive protein and other inflammatory markers may be present; but are non-specific.

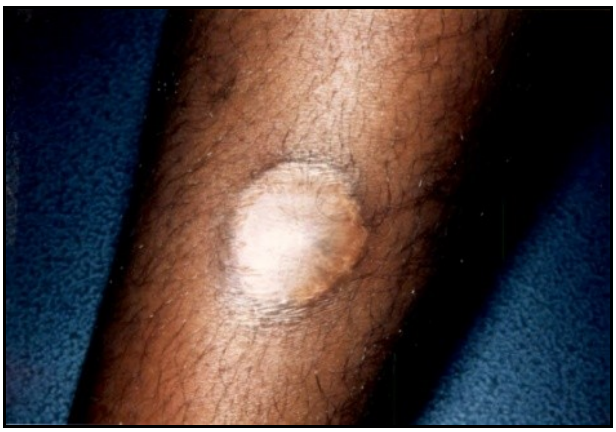

Fig 5. *Characteristic parchment-paper-like thin depigmented scar of healed Tropical Phagedenic Ulcer (From MD thesis of Beverley Adriaans submitted to the University of Cape Town, South Africa, 1988).*

Table 2. Differential diagnosis of Tropical Phagedenic Ulcer

Differential diagnosis	**Similarities with TPU**	**Differentiating features**
Ecthyma (Pyoderma gangrenosum)	Antecedent minor injuries Lower limb distribution Dirty granulation tissue	Crust formation Superficial ulcer of small size Quick healing with treatment Isolation of Gram positive cocci Absence of epithelial hyperplasia
Cutaneous diphtheria (Castellani's 'tropicaloid' ulcer)	Presence of pseudomembrane Onset as vesicle Overhanging edges of ulcer Punched-out margins of ulcer	Multiplicity of ulcers Isolation of *Corynebacterium* Irregular shape of ulcers Distribution in upper part of body
Cutaneous Leishmaniasis	Onset as papule and vesicle	Crust formation Distribution in trunk, arms and face Irregular shape of ulcer Demonstration of Donovan bodies
Necrotizing fasciitis (e.g. Noma, Meleney's ulcers, Fournier's gangrene)	Rapidly spreading necrosis Fetid smell Malnourished patients Tropical prevalence	Different anatomical distribution Absence of sclerotic chronic phase
Sickle cell ulcers	High incidence in second decade Anatomical distribution in legs Painful lesions	Onset in deeper tissue rather than skin Different geographical distribution Poor response to antibiotics Presence of abnormal HbS / sickling of RBC Isolation of aerobic bacteria
Buruli ulcer	Pediatric preponderance Undermined edges Identical geographic distribution	Subcutaneous rather than epidermal onset Painless ulcers Absence of papules or pustules Irregular shape and huge size Demonstration of mycobacterium Poor neutrophilic infiltration More Extensive collagen destruction
Yaws	Pediatric Prevalence Anatomical distribution in legs Punched out circular ulcers	Multiplicity of ulcers Absence of Fusobacterial isolates
Bazin ulcer (Erythema induratum)	Distribution in legs Punched out edges	Presence of vasculitis Painless, small, deep ulcers
Venous ulcers	Distribution in ankles	Rarity in children Painless, Irregular, superficial ulcers Associated with varicose vein
Cutaneous blastomycosis	Phagedenic spread of ulcers Indolent nature	Different anatomical distribution Demonstration of fungus Quick response to antifungal treatment

DIFFERENTIAL DIAGNOSIS

Several tropical diseases clinically mimic the skin lesion of TPU. (Table 2; Fig. 6) Radiographic differential diagnoses of bone involvement in TPU include Brodie's abscess, osteomyelitis, Ewing's sarcoma and osteosarcoma.[59,67] The absence of a *penumbra sign* in the bone MRI is said to differentiate TPU changes from osteomyelitis.[11]

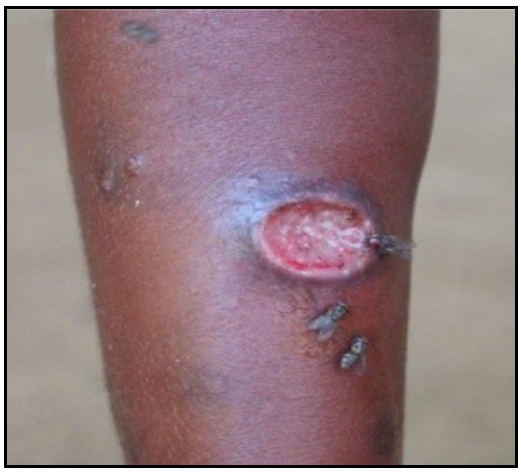

Fig 6. *Characteristic leg ulcer of Yaws mimicking Tropical Phagedenic Ulcer. (Reproduced from Mitja O et.al, PLOS Neglected Tropical Diseases 2017; DOI:10.1371/journal.pntd.0005136 under Creative Commons Attribution License)*

TREATMENT

Blaine[5] summarized the principles of treatment by the acronym ACEER: **A**rrest of the spread of ulceration, **C**learing of infection, **E**arly and adequate separation of pseudomembrane, **E**ncouragement of granulation tissue formation and promotion of **R**e-epithelization.

Marsh and Wilson [42] recommended a "lock-up treatment" wherein the affected limb is immobilized it in a plaster-of-Paris cast. Advantages of immobilization and closed dressing include protection of growing capillaries of granulation tissue and delicate epithelium from mechanical damage, prevention of tissue desiccation, reduction of pain, avoidance of further auto-inoculation, prevention of environmental contamination and promotion of lymphatic stagnation that helps in halting the systemic spread of infection.[42] Marsh and Wilson claimed that 59 of the 85 cases treated with lock-up therapy completely healed within 2 weeks. But, Yesudian and Thambiah [40] disapproved any form of closed dressing because it creates a warm moist anaerobic environment that is conducive to the growth of pathogens. It is to be noted the Marsh and Wilson used topical antimicrobials in conjunction with 'lock-up'.

Several topical and systemic agents have been used in the treatment of TPU. (Box 4 &5) Some of them are obviously dangerous, while others are of doubtful use. For example, mercuric perchlorate, tar, hydrochloric acid and formalin can no longer be considered as ethical treatments. Some agents are useful selectively in specific cases. Fish oil dressing is shown to remove malodor and to promote epithelization in chronic ulcers but not in acute ulcers.[48] In fact, oily dressings may worsen acute infections. Therefore, the treatment of TPU should be tailored to the stage of disease and to the patients' condition.

Acute Phase Management

In early stages, systemic antibiotics and frequent washing of wounds are indicated. The antibiotics should cover gram positives, gram negatives and anaerobes. Ideally, a combination treatment with third-generation cephalosporin, penicillin, erythro mycin, streptomycin, co-amoxiclav, metronidazole and aminoglycosides is recommended. Although clindamycin has a good anaerobic coverage, it should be avoided because FU are genetically resistant to it. In multidrug resistant cases, rifamycin or oral tetracycline may be used in older children with due precaution. Systemic antibiotics are useful only in acute stage, but not in chronic stages.[87]

The best topical treatment is perhaps, frequent washing of the wound with soap and running

water. Hydrogen peroxide, boric acid, povidone-iodine and potassium permanganate are also useful. Hydrogen peroxide and digestive enzymes (e.g. chymotrypsin) may be useful in the separation of the pseudomembrane. Epithelial irritants (e.g. metronidazole), oil-based applications (e.g. Vaseline) and agents that can provoke bleeding (e.g. streptokinase) are better avoided in the acute phase.

Topical antibiotics (e.g. gentamicin, penicillin) are not recommended as they facilitate the emergence of resistant strains. As the offending bacteria are deep inside the wound, topical applications are mainly useful in controlling the secondary pathogens rather than the FU. Hyaluronidase is claimed to improve the penetration of topical agents.[5] Silver sulphadiazine and silver nitrate are ideal topical agents.

Chronic Phase Management

In the chronic stage, when infection is no longer a threat, protection of growing epithelium by oil-based dressings or 'lock-up' therapy is preferable. Topical and systemic antibiotics are of limited use in indolent ulcers. Management of complications and rehabilitation of crippling limb deformities are inherent components of long-term care. When the underlying tendons and bones are damaged, through surgical debridement is essential for healing.

Role of Surgery

Surgical debridement is best avoided in the acute phagedenic phase as it may precipitate septicemia. However, periodic gentle debridement is indicated in the case of extensively deep ulcers.

The importance of surgical excision in the healing of chronic ulcers was appreciated as early as in 1930s.[54,65] It is hypothesized that excision of the ulcer removes the source of cytotoxic chemicals that inhibit natural healing.

Surgical intervention may be (i) curettage of ulcer bed with or without skin grafting, (ii) tangential excision of ulcer with immediate or delayed skin grafting, (iii) excision of ulcer with primary suturing of flaps and (iv) allograft or xenograft used as temporary dressing materials. The senior author (VR) has used collagen dressing with good results.

Autologous graft may be Thiersch partial thickness sheet-graft or Reverdin's pinch graft (also known as seed or punch graft). Seed grafts give better results than sheet grafts and are implementable by trained paramedical workers in remote African villages.[65,72] Graft failure is recorded in 2-30% especially when applied over the tibia.[54,65] In such cases cortical drilling is recommended to encourage sprouting of granulation tissue from the medulla.

Newer Treatments

Smith suggested a 'cocktail therapy' that includes antimicrobials (antifungal if necessary), platelet-activating-factor (PAF) antagonists and hyperoxygenating agents (e.g. hydrogen peroxide).[61] H_20_2 not only kills microbes by liberating nascent oxygen but also deactivates PAF by oxidation. It is to be noted that this 'cocktail therapy' does not appear to have been tested in clinical patients.

Recently, herbs used by the Apsokok nomadic tribe of Papua New Guinea have been found to be useful in healing TPU.[9] *In vitro*, they inhibited *Staphylococcus*, but not FU. The active ingredient of *Homalium foetidum* (known locally as Malas) extract that exhibits antibacterial property has been identified as coumaroylquinic acids. Extracts of *Alstonia scholaris* (Rambaka) and *Pangium edule* (Kali) were found to stimulate collagen synthesis by the fibroblasts and inhibit MMP-9, a proteolytic enzyme that destroys growth factors and extracellular matrix. Burnt ashes of these plants are applied to the TPU. Possibly, it acts as an activated charcoal dressing, that absorbs toxins and promotes wound healing.[9]

Box 4. Topical agents used in the treatment of Tropical Phagedenic Ulcer

Topical agent	Reference *
Acriflavine‡	James 1938
Arsphenamine [Salvarsan]‡	Heath1924
Aureomycin [Chlor-tetracycline]	Lasbrey 1952
Azochloramid	Marsh 1948
Bacitracin	Blaine 1958
Banana leaf dressing‡	Adriaans 1988
BIPP‡	Marsh 1945
Boric acid	Brienl 1915
Calcium chloride‡	Loewenthal 1963
Carbolic acid‡	Das 1952
Cetyl pryidinium bromide‡	Blaine 1958
Chincona power‡	Innes 1931
Chloramphenicol	Payne 1951
Chymotrypsin	Blaine 1958
Cod liver oil	Corkill 1939
Copper sulphate‡	Gunther 1938
Diluted hydrochloric acid‡	Adriaans 1988
Eucalyptus oil	Patterson 1908
Eusol	Manson-Bahr 1936
Extract of manchineel tree‡	Adriaans 1988
Formalin‡	Boucher 1916,
Gentamicin	Adriaans 1988
Gentian violet	-
Hyaluronidase	Blaine 1958
Hydrogen peroxide	Ziprkowski 1955
Hyperbaric oxygen	Ledingham 1975
Hypotonic saline dressing	Clement 1936
Linseed oil	Marsh 1948
Mercuric perchlorate‡	Sen 1922
Metronidazole	Lindner1968,
$MgSO_4$-Glycerine paste	Earle 1942
Neomycin	Blaine 1958
Olive oil	Marsh 1948
Oxy-tetracycline	Ampofo 1951
Papaya pulp‡	Colombetti 1965
Para-chloro-meta-xylenol	Marsh 1948
Paraffin gauze	Gupta 2002
Penicillin	Drasar1987
Polymyxin B	Blaine 1958
Potassium permanganate	Heard 1908
Povidone iodine	Adriaans 1988
Quinine sulphate/ Cinchona‡	Innes 1931
Radiation therapy locally‡	Clement 1936
Saline (Physiological)	Marsh 1948
Silver nitrate	Marsh 1945
Silver sulphadiazine	Adriaans 1988
Sodium bicarbonate	Innes 1931
Streptokinase	Blaine 1958
Streptomycin	Gupta 2002
Sulphathiazole	Marsh 1948
Tar ‡	Adriaans 1988
Trichlorophenylmethliodosalicyl	Marsh 1945
Vaseline	Patterson 1908
Vincent's powder†	Corpus 1924
Zinc oxide	Marsh 1948
ZIPP‡	Connell 1933

** References need not necessarily indicate the first known usage. Detailed bibliography (not included herein) is available from the authors on personal request.*

†It contains l part sodium hypochlorite and 9 parts boric acid.

‡ These are merely of historical interest. They should never be used in modern medicine.

BIPP (Bismuth, Iodoform Paraffin Paste)
ZIPP (Zinc oxide, Iodoform Paraffin Paste)

Box 5. Systemic agents used in the treatment of Tropical Phagedenic Ulcer

Systemic drug	Reference*
Aureomycin [Chlor-tetracycline]†	Ampofo 1950
Cacium chloride†	James 1938
Chloramphenicol	Payne 1951
Gentamicin	Adriaans 1988
Metronidazole	Lindner1968,
Neosalvarsan†	Apostolides 1922
Oxytetracycline	Ampofo 1951
Penicillin	Webb 1946
Streptomycin	Gupta 2002
Sulphonamides	Earle 1942

** References need not necessarily indicate the first known usage. Detailed bibliography (not included herein) is available from the authors on personal request.*

†No longer in use

COMPLICATIONS

Several complications of TPU have been documented in the literature, all of which are rare in modern days. Early medical intervention and the availability of better antimicrobials may be the reason for this change.

Squamous Cell Carcinoma

Malignant transformation occurred in 2-9% of TPU after a mean lapse of 3 years.[2,71] Fortunately, malignant cells do not metastasize thanks to dense fibrosis and poor vascularity of the underlying soft tissues. This is similar to Marjolin's ulcers.[59]

Osteitis

The tibia, due to subcutaneous location, is vulnerable to secondary changes in TPU.[59,67] Bone just beneath the ulcer is usually involved; however, remote bones are rarely affected. The pathogenic sequence of bony lesions[59,67] can be summarized as follows:

1. Lifting of the periosteum due to inflammatory edema leads to periosteal reaction and new bone formation. This causes a fusiform or 'sunburst' appearance in radiographs.
2. The involucrum blends with the existing cortex resulting in an 'ivary-osetoma like' cortical sclerosis. This is seen as periosteal heaping or thickening in radiographs. Rarely an onion-peel appearance is resulted. The new bone may be as thick as 2 cm[59] and is called as 'ulcer osteoma'.[67] Two different types of ulcer osetomas are known to occur. The cancellous variety is common in the fibula while the sclerotic type is common in the tibia.[67] Sclerotic osteomas are reversible by early intervention while the cancellous osteomas almost invariably require surgical excision.[67]
3. Irregular new bone formation causes undulations in the bone cortex.
4. The fragile involucrum disintegrates leaving behind a sequestrum.
5. Separation of the sequestrum leaves behind a saucer-shaped crater (*cortical ulcer*), thus weakening the bone cortex
6. The weakened bone, together with inflammatory epiphyseal fusion and fibrosis of adjacent soft-tissues causes bending of limbs with a convexity towards the ulcer site.[59] Bowing of the tibia, valgus deformity of the ankle and drawing together of the tibia and fibula are well known.
7. Two types of medullary bone changes occur in TPU: (i) Osteoporosis-like change occurs distal to the ulcer site due to disuse atrophy; (ii) Osteomyelitis-like change occurs at the ulcer site due to inflammation effect.
8. Deformed, osteoporotic bones are prone for pathological fractures

In addition to bony changes adjoining tendons and muscles may also be involved in necrosis and fibrosis, thus adding to deformity and disability.

Hepatitis-B

About 26% of TPU patients were found to have associated hepatitis B in Kiribati and Gambia.[63,64]. The ulcer exudates were positive for Hepatitis virus. Thus, open TPU are a source of great public health concern.

Rare Complications

Gas gangrene,[68] tetanus,[68] thrombophlebitis, toxemia and death are now not seen. Psychological depression due to non-healing ulcers has not been adequately studied.[54]

PROGNOSIS

The mean healing time of ulcers is 4-6 months. Recurrent ulcers are not unknown in TPU. They occur at the same site in 25% and at a different site in 65% cases.[2,59] The morbidity of bone involve-ment and scar contractures are not well studied. Amputations were common in the pre-antibiotic era.[58] In 1972, out of the 230 cases of

TPU, 11 patients required amputation of a limb, among whom 7 had squamous cell carcinoma.[71] There were no deaths reported in the last 70 years!

PREVENTION

The importance of good nutrition, a clean environment and personal hygiene cannot be overemphasized. Protective trousers and footwear, applying oil to legs during outdoor playing and files control measures are recommended to prevent the spread of infection. Daily bathing by applying soap is perhaps, the single most important preventive strategy of TPU.

Epilogue

More than 80% of the literature on TPU is at least 4 decades old. Almost all of them are descriptive observational studies. Randomized controlled trials and meta-analysis are almost nonexistent. Hence recommendations made in this review are of level-4 evidence. This review calls for more robust scientific studies on TPU.

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Patient Perspective

Born Unable To Swallow

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Keywords
Esophageal atresia
Congenital malformations
Patient perspectives

Abstract

Mr. Stephen Wyles was born with esophageal atresia. He is running a voluntary patient support group (Birth-defect.org) that helps children born with esophageal atresia in resource poor tropical countries. In this article he shares his perspectives and ordeals of a child born with esophageal atresia.

I was born in 1962 in England with a serious condition called esophageal atresia (EA) with tracheo-esophageal fistula (TOF).[1] My journey with this medical condition has been challenging, scary and complicated. It was a learning experience and reward that has given me an opportunity, especially in the latter years, to meet many different people from many different parts of the world. I would like to take this opportunity to thank all the doctors, nurses, allied health professionals and support staff, many of whom are behind the scenes. They have treated and looked after me over the last 62 years.

When I was conceived, my mother's belly became very large. My father would joke that he could see her tummy coming round the corner before he could actually see her. In those days people were not so aware of polyhydramnios and its connection with babies with EA. So, my mother did not have arrangements to be in a hospital when she gave birth to me. I was born in a small village in Hampshire England. It was noticed immediately that I was choking on secretions and I was rushed to Southampton General Hospital, where I had an operation that saved my life. I remained in the special care baby unit for many weeks, whilst my condition was stabilized.

My life as a child was a struggle at times, to say the least. No one really understood the condition I was born with. Outwardly, I also looked normal and cured; but my esophagus was very narrow which meant I could only eat certain things and my esophagus needed stretching over the years as I grew. I also developed a 'TOF cough' (a characteristic loud, barking cough). Hospital stays were hard, in those days. I was left in hospital alone; my parents were not allowed to stay with me.

When I attended nursery and primary school, teachers and assistants, did not understand how to treat me if I got food stuck in my esophagus. I remember clearly a lady forcing me to have white bread and water to help remove the blockage, which only made it worse. To date I hate being in cold water; I think this is due to one of my hospital memories as a child. They put me in a cast-iron

bath with salt water and made me to stay there till the water went cold. I also have a fear of needles and faint at the sight of them. These are all due to the treatments I had had as a child, which my body now associates with trauma and blacks out.

I still have a large operative scar on the side of my chest. I used to pretend at school that I had been attacked by a shark. I had many junior doctors examining my scar and been taught about my condition. A doctor who examined me explained that my scar had been done really neatly and that I had been lucky to have had the surgeons I did at that time. I used to know their names previously; but only remember one name now, Dr Roundtree, to whom I am very grateful. The hospital has since deleted all my records which I would like to see now. Today people are able to keep their own records thanks to the advancements in digital technology.

When I was a young adult, my mom did a drawing of the procedure that I had undergone. (Fig 1) With this she explained to me the rare condition that I suffered. It helped me to explain my problem if I need to be admitted to hospital or to see a doctor. Once I developed an ulcer in the esophagus and was vomiting blood. Doctors assumed that I was an alcoholic and had acquired the ulcer through drinking too much. They were amazed when I said that I was teetotaler and produced the drawing.

At another time, in my 20's, a young doctor dismissed my condition when she knew my age. She explained that no baby survived with TOF stricture in 1962, and discharged me after having watched me drinking sips of water. She was not even convinced by my scar. Two days later I returned to the hospital, and was greeted by the same doctor. It was then 4 days since I had managed to swallow any food. I was admitted and after 3 days of being in the ward, I was sent for a barium meal study. I invited the skeptical doctor to come with me. Standing between two large plates of metal, I swallowed the white chalky substance; and all that I heard from other side was the young doctor gasping in shock at what she was seeing - a disc of chestnut with barium sliding over it in a very thin esophagus. After 10 days of my suffering without any food, she finally believed me.

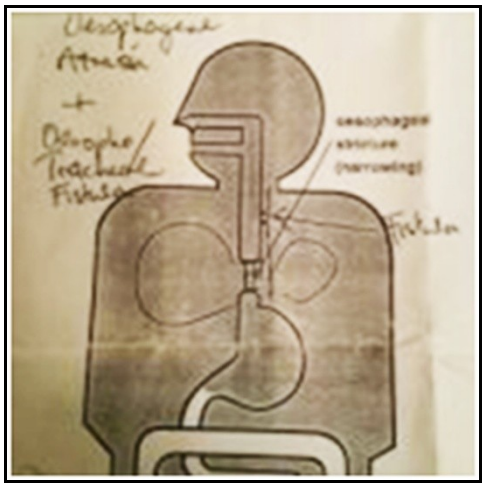

Fig 1 *Hand drawn sketch of Mom*

Every day whilst eating food it sticks going down; but I have learnt to clear it using my own techniques that seem natural to me. I help parents of TOF babies and explain that they should not panic if their children get the food stuck. They should just encourage the children to find their own ways of moving it down. Parents should reassure that it is normal to them and not something to panic about.

In 2013, I developed a website (www.birth-defect.org) to help parents of children born with TOF as I felt that there were not enough parental information materials on this condition. I also have a Facebook site and I cannot just believe as to how this has grown. I am now in communication with lots of surgeons around the world especially the resource poor tropical countries and I could provide support to parents of babies born with this condition all around the world. I help to connect parents with surgeons and assist parents to lend support to other parents of similarly

affected children by connecting them through the Facebook page.

On one occasion I was mentioned in the American News for connecting a family with a surgeon, which is amazing as I live in the UK. I have received so many grateful messages from parents. I even received an award from the Royal Family of UK, which I'm very proud of. I am also a co-author of two medical papers.[2,3] Who knew that I would get my name on a medical research paper!

In collaboration with a top research hospital in the USA, I developed an idea of making a worldwide survey of the condition. Unfortunately, due to various issues it was not as successful as I had hoped, but my dream would be to work with all the pediatric surgeons and charity groups across the world to try and achieve this. One day I hope we can all be involved in creating a world survey again. My website hits 193 countries last year, so I know we can do it.

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Letters to the Editor

Double-J Stent Removal

Dear editor,

I read with great interest the article on non-invasive removal of double-J (DJ) stent.[1] It is always a welcome to remove DJ stent in children without anesthesia and cystoscopy. However, I wish make some pertinent observations regarding the paper.

Firstly, this technique described by authors is not new. As early as 1986, Siegel *et.al* developed a similar technique of stent retrieval using a mono-filament nylon extraction string which was tied to the distal end of the DJ stent and its outer end taped to the penis or abdomen.[2] The technique has subsequently been modified by several other researchers.[3,4] The authors also completely ignored citing several large randomized controlled studies and systematic reviews that are relevant to the paper.[5,6] Further, authors should have also clearly stated as to what was their modification and as to how was it better than the other existing techniques of string-based double-J stent removal. Secondly, the authors should have discussed the advantages and disadvantages of their technique in comparison to the alternative techniques such as the Vellore technique.[7] Thirdly, spontaneous expulsion of the free end of the extraction string is a possibility rather than a certainty. It would be interesting to know, in how many of their patients the authors faced this problem of non expulsion of the extraction thread in urine stream. Fourthly, once expelled in urine stream, how did the authors prevented premature accidental pulling of the thread by the child or by his parents? Accidental dislodgement is as high as 24% in one study.[8] Fifthly, in case of bilateral stents that require removal at different dates, how did the authors manage to identify the side of stent that is to be removed? Sixthly, did the authors encounter spontaneous knotting of thread inside bladder? Seventhly, the silk string is prone for infections owing to its braided nature. This is especially true when the string is hanging outside.[9] It may also introduce ascending infection. Did the authors consider using any other monofilament strings to reduce the risk of infection? Did they use prophy-lactic antibiotics? What was the rate of UTI in the authors' series? Finally, the practical utility of the article would have been enhanced had the authors added to their discussion about newer stents such as the bio-degradable stents[10] and the magnetic tipped stents[11] which render their removal either unnecessary or easy respectively.

Notwithstanding these critical observations, I would like to congratulate the author for having given a new perspective to the removal of DJ stent in children without anesthesia or cystoscopy.

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AUTHORS' REPLY

Dear editor,

I read with great interest the long critical letter by Dr. Ijaz Ahmad. I appreciate the efforts taken by him to review the literature. Some of his points are really interesting to read. However, we wish to make the following observations:

1. All the studies cited by him have been done in adults. Ours is exclusively done in children.
2. Braided threads causing infection is not a significant issue. Level-1 evidence comparing monofilament versus braided suture in the urinary tract is not available. When a larger foreign body (stent) itself is left inside the bladder, a smaller thread ought not to be a cause of great concern.
3. Bilateral stents are usually removed at the same time. Whether the right or the left stent comes out first is immaterial.
4. For babies on diapers, accidental premature removal is unlikely.
5. This is the first study of its kind from India in the pediatric age group. General anesthesia and cystoscopy for stent removal in this setting is always difficult. If it can be avoided, so much the better.
6. If the extraction string does not emerge spontaneously, all is not lost; it can always be pulled out by the conventional cystoscopic method.
7. Other modern stents mentioned by Dr Ahmed are too costly and we do not want to escalate the cost of care as ours is a charitable hospital.

Thank you for the opportunity to respond to the criticisms.

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Announcement

Dear Scholar

Editors of Pediatric Surgery in Tropics takes immense pleasure in inviting you to this Reviewer Training Workshop. It is intended to train potential journal reviewer in critical appraisal of scientific manuscripts and providing standardized review comments. At the end of the course participating Pediatric Surgeons will be recognized as Certified Reviewers of the journal. All participants will receive CME credits points as granted by National Medical Commission of India.

Delegate Fee: None; But prior registration is must
For Contacts: Dr. Krishnakumar G 8903860378
Dr. Aravindh R 9003548950
Dr. Raveenthiran V 9443310182

Organizing Chairman: ***Dr Vivek Gharpure***
Organizing Co-Chairmen: ***Dr Raveenthiran***
Dr Yogesh Kumar Sarin
Organizing Secretaries: ***Dr. Krishnakumar Govindarajan***
Dr. Aravindh Radhakrishnan

Guest Faculty

Dr. Rakesh Agaarwal
Director, JIPMER, Pudhucherry
Editor, Indian Journal of Gastroenterology and Hepatology
President, World Association of Medical Editors

Dr. Peush Sahni
Editor, National Medical Journal
Past-President, World Association of Medical Editors

Dr. Yogesh Kumar Sarin
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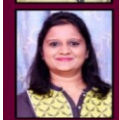
Past-President, Indian Association of Pediatric Surgeons
Emeritus Editor-in-Chief, Journal of Neonatal Surgery

Dr. Devendra Mishra
Executive Editor, Indian Pediatrics

Dr. Vivek Gharpure
Chief Editor, Pediatric Surgery in Tropics



Dr. Aarti Garg
Editor, Journal of Clinical and Diagnostic Research

Dr. V. Raveenthiran
Editor, Pediatric Surgery in Tropics
Former Associate Editor, Indian Journal of Surgery
Former Editor-in-chief, Journal of International Medical Sciences Association
Former Editor, Journal of Neonatal Surgery

Program Schedule

Tentative program on 14 July 2024
Time 9.30 am to 1 pm (Indian standard time)
Meeting Code: Will be informed personally to registered participants

Time	Topic	Speaker
Talk 9.30 - 9.50 Discussion 10.00	Introduction to peer review process - Definition, work-flow pattern, benefits, importance of peer reviewing	Dr Vivek Gharpure
Talk 10.00-10.20 Discussion 10.30	Critical reading of a manuscript - Assessment of scientific contents	Dr Rakesh Agaarwal
Talk 10.30 - 10.50 Discussion 11.00	Detecting research misconduct (including plagiarism and figure manipulations)	Dr Aarti Garg
Talk 11.00 -11.20 Discussion 11.30	Writing review comments (including language of the reviewer, and dealing with revisions)	Dr. Peush Sahni
Talk 11.30-11.50 Discussion 12.00	Ethics of peer review - Reviewer behavior, Reviewer COI	Dr Devendra Mishra
Talk 12.00-12.20 Discussion 12.30	Assessment of Titles, keywords, abstracts, statistics, figures and tables	Dr. Yogesh Sarin
Talk 12.30 - 12.50 Discussion 1.00	Assessment of language and style of presentation including IMRAD and guidelines	Dr V. Raveenthiran