SEPA Based Flap for Covering Defect after Adult Exstrophy Epispadias Repair

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ABSTRACT

INTRODUCTION
Bladder exstrophy is a complex, rare disorder that affects the foetus in utero. Here the abdominal wall is not fully formed, leaving the pubic bones separated and the bladder exposed in the lower abdominal wall.

METHODOLOGY
Single-stage reconstructive and repair surgery was undertaken in adults diagnosed with bladder exstrophy epispadias complex. The superficial external pudendal artery flap was considered for covering the defect after extensive reviews and discussion.

CONCLUSION
An uneventful surgery with recovery and a two-year follow-up post-surgery did not account for any complications following the procedure. This shows that the superficial external pudendal artery flap can be considered a favourable option for urologists in the repair of bladder exstrophy epispadias complex when undertaken in a single sitting surgery, especially in adults.

KEYWORDS
Adult Bladder Exstrophy, Epispadias Repair, SEPA (Superficial External Pudendal Artery), Axial-Pattern Flap, Single-Stage Exstrophy Epispadias Repair.

MeSH Terms
Bladder Exstrophy, epispadias repair, superficial external pudendal artery, axial-pattern flap.
INTRODUCTION

Bladder exstrophy is a rare congenital malformation in which the anterior wall of the bladder is absent because of which the posterior wall is exposed externally.[1] Commonly occurring as bladder exstrophy epispadias complex (BEEC), it has a variable expression with a wide range of anomalies associated with the abdominal wall, pelvis, urinary tract, genitalia, spine and even the anus, in addition to an anterior midline defect.[2] There is a failure of fusion in utero of the infraumbilical abdominal wall, the anterior wall of the bladder with a split urethra and splayed out genitalia and musculature along with pubic diastasis. The exact embryopathogenesis of bladder exstrophy is unknown. It usually is an incidental finding during routine pregnancy in ultrasound confirmed with MRI.[3] Though rare it affects the quality of life of both the patient and their families. As the bladder is unable to hold the urine, urinary incontinence is troublesome apart from associated sexual dysfunction and increased risk of bladder cancer.

Surgical manipulation is the only treatment. However, the success of the surgery depends on the severity of the defect. To prevent its associated symptoms and to improve the quality of life, multiple major reconstructive surgeries are usually undertaken within months of the birth of the child. There are long-term functional, psychological and social outcomes in children undergoing repeated hospital admissions and extensive multiple operations. Issues include long-term adjustment problems in relation to incontinence, ambulation, psychological disturbances, sexual dysfunction, self-esteem and social integration.[4] Adult bladder exstrophy results in a lack of paediatric surgical expertise, paediatric anaesthesia and paediatric ICU which are common hurdles in developing countries. The situation worsens amongst the rural population that doesn't have access to basic health care itself. Poor surgical outcome and ignorance of the condition compound the situation further.

We had eight cases of adult bladder exstrophy who came in for surgical treatment. As affordability was a consideration along with drastic improvement in their condition, a single-stage reconstructive surgical approach was taken. Based on the anatomical positioning of the defect, the closest collateral blood supply was selected based on numerous discussions and work experience with plastic surgeons. An axial-pattern flap using the superficial external pudendal vessels was undertaken to cover the anatomical defect in patients undergoing extrophy and epispadias repair surgery.

METHODOLOGY

Eight cases of adult exstrophy epispadias were operated upon from January 2013 to June 2016. Among them 7 were males and one was female. The average age of presentation was around 18 years. The patients were evaluated by clinical examination, Ultrasound KUB and X-Ray KUB.

Reconstructive surgery was planned, to be executed in a single stage. Upon consent from the patient for the same, surgery was undertaken. The patients were monitored throughout the surgery with no untoward events occurring on the table. The immediate and delayed postop period was also uneventful. The patients were discharged after 14 days of hospital stay with subsequent follow-up at 3 weeks, 6 weeks initially and thereafter every 3 months till 2 years following the surgery. There were no complications till the end of the follow-up period post-surgery.

DISCUSSION

Bladder exstrophy reconstruction was first advocated by Jeffs and Cendron as early as the 1970s. The staged approach has undergone many significant changes with continuous modification and improvements.[5] As many components of the defect have to be corrected, understanding the magnitude and the consequences of the same is essential.

Soft Tissue Anomaly

Clinical examination showed open bladder defects involving the anterior wall of the bladder and the anterior abdominal wall. Associated defects include bilateral ureteral orifices opening into the anterior abdominal wall defect with the urine dribbling externally (Figure 1.). Associated anomalies include absent umbilicus, penopubic epispadias and undescended testes in males.[6] When the bladder is small, fibrosed, inelastic and covered with polyps, functional repair may be impossible.

Skeletal Defects

Classic bladder exstrophy has a mean external rotation of the posterior aspect of the pelvis at 12 degrees on each side, retroversion of the acetabulum and a mean of 18 degrees external rotation of the anterior pelvis along with 30% shortening of the pubic rami. This is in addition to the diastasis of the symphysis pubis.

Abdominal Wall Defects

The triangular defect caused by the premature rupture of the abnormal cloacal membrane is occupied by the exstrophy bladder and posterior urethra. The fascial defect is limited inferiorly by the intrasymphysisal band which represents the
divergent urogenital diaphragm. This band connects the bladder neck and posterior urethra to the pubic ramus an anatomic study. The anterior sheath of the rectus muscle has a fanlike extension behind the urethra and bladder neck that inserts into the intrasymphyseal band.[4] Investigations using MRI into the relationship of the rectus muscle and fascia to the urogenital diaphragm are a must before undertaking the surgery. This is a must for the soft tissue assessment of the pelvic floor which has to be reconstructed.

The superficial external pudendal artery (SEPA) flap is very versatile and has been used in other defect repair surgeries.[7] This fasciocutaneous flap used in these single-stage reconstructive surgeries is very mobile and can be rotated almost 180 degrees on either side. There are three superficial branches of the femoral artery in the groin. They are the superficial circumflex iliac artery (SCIA), the superficial inferior epigastric artery (SIEA) and the superficial external pudendal artery (SEPA). All three vessels radiate out from the saphenous opening and give off branches which are mostly arranged radially, like the spokes of a wheel, centred on the saphenous opening.[8,9,10] As the SEPA had remained unexploited, the present study was undertaken to ascertain its potential use in the closure of adult exstrophy and epispadias repair on either side of the midline, extending from the symphysis pubis to the umbilicus, designing the defect. Accurate knowledge of the anatomy of the vessel concerned is a prerequisite for skin flap (Figure 2).

**Point of Technique**

All eight cases were operated upon by a single surgeon. The procedure performed was single-stage exstrophy epispadias repair followed by SEPA based flap cover. The mean operating time was 8 hours. The steps included 1) Mobilization of the Bladder Plate 2) Closure of the Bladder Plate 3) Augmentation Ileocystoplasty 4) Cantwell-Ransley epispadias repair 5) SEPA flap and closure of the abdominal wall defect and 6) Covering of raw abdominal area with a skin graft.

**CONCLUSIONS**

Adult bladder extrophy is a traumatic condition to live with. Apart from functional or a disability loss there is more emotional stress affecting the quality of life for the patient. In western countries and advanced centres, extrophy of the bladder is operated in neonate age groups and primary closure of the abdominal wall is achieved with soft tissue mobilization and osteotomies. However, in adults, it is not possible to primarily close the abdominal wall given the non-malleable nature of the bones at that age. Hence we use SEPA based flap which is a reliable, convenient and versatile flap giving good anatomical closure with excellent functional and cosmetic results (Figures 3 & 4.). It is easy even for the urologist to do the flap surgery.

**REFERENCES**