

Bladder exstrophy spica new design

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ABSTRACT

Introduction: Bladder exstrophy such a congenital malformation of musculoskeletal, urogenital systems extending superficially to the anterior abdominal wall with the bladder being exposed, with incidence of 1:30000 in classic bladder exstrophy

Methods: 18 children were included in this case series, 21 osteotomy assisted closure utilizing Sponseller's osteotomy were done, then immobilized in newly designed -KZ- spica cast for 4-6 weeks period.

Results: 18 patients who underwent 21 osteotomy assisted closure of exstrophy, 7 girls 33.33% and 14 boy 66.67% (chart 1), age of patients at time of intervention varied from 3 months to the age of 25 months, follow up period up to 3 years in some cases, on 14th day post-operatively ensuring complete closure of urological wound then at 4 and 6 weeks for cast removal after bony union

Spica cast success was to achieve bony healing in appropriate time of 4-6 weeks post-osteotomy without delayed or non-union, Success rate of spica cast to achieve bony healing was 100%, with minimal skin rash as a complication

Keywords: Bladder, design, spica, exstrophy

INTRODUCTION

Bladder exstrophy is a disease including genitourinary-musculoskeletal-intestinal congenital malformation resulting from premature rupture of cloacal membrane rather than its persistence leading to epispadias, classic bladder exstrophy (CBE) and cloacal exstrophy (CE) according to severity and systems involved with incidence 1:20000 in CBE up to 1:150000 in CE, male predominance over girls 2:1. (Harris et al., 2024)

Reinsertion of the bladder and repair require tension free closure of the anterior abdominal wall and pelvic bones with the aid of osteotomies, being first done and adopted by Trendelenburg over the years numerous osteotomy types were used -posterior, anterior and oblique-, Sponseller's osteotomy is adopted in our pediatric orthopedics unit in a North African university tertiary hospital -Mansoura University- to be the formal exstrophy osteotomy carried out by performing two perpendicular limbs, horizontal cut like the formal Salter's pelvic osteotomy and vertical cuts being parallel and 1 cm lateral to the sacroiliac joint performed as a closing wedge of the inner plate alone while commencing greenstick fracture to the outer table with exerting inward force up on the iliac wings, it was 1st described on February 1996, and hasn't gain popularity in Egypt and North-African area yet. (Gearhart et al., 1996)

The pelvic osteotomy necessitate stabilizing method which supplies the pelvis with stability necessary for both bony healing, pain control, and finally ensuring the position of bony pelvis required for tension free nature of the closure with secure symphyseal apposition even bringing pelvic floor muscles into more anatomical position, using hip spica cast with special positioning of the lower limbs or with external fixator, first means of immobilization was described and adopted by Trendelenburg also following pelvic osteotomies to immobilize osteotomy and approximate pubic bones, he made a device called it "pelvic sling" (Haney et al., 2022, Pathak et al., 2020)

Aim of the study

Assessment of new design of bladder exstrophy hip spica applied success and failure in bony support to achieve healing.

Assessment of newly designed exstrophy cast complications

Methods

This research had Institutional Review Board (IRB) approval under the approval number/ID MD.21.08.505, all participants parent gave written informed consent to participate in the study.

In this case series, it was done in a specialized pediatric orthopedic unit in collaboration with a pediatric urology unit at a North African tertiary university hospital -Mansoura university, Egypt-.

Sixty-four children were identified in this retro-prospective study in which seventy-four pelvic-osteotomy assisted exstrophy closure were done, then after exclusion of 9 cases who were put in external fixator post-operatively, forty-four cases in old formal spica cast, twenty-one cases were included in this study who were immobilized in newly designed hip spica post Sponseller's osteotomy assisted exstrophy closure.

Inclusion criteria

All neonates and pediatric group since birth presented with bladder exstrophy, and were immobilized in the newly designed hip spica post-operatively.

Exclusion criteria

1. Any neonates aged 72 hours or less which can be managed with cast without osteotomy
2. Cases who were put in external fixator
3. Cases who were put in formal old hip spica
4. Any case in which clinically with aid of examination under anesthesia while positioning both lower limbs in adduction to their body midline then maximum femoral internal rotation could be obtained if with this means the pubic diastasis is eliminated completely and getting symphyseal diastasis completely closed and eliminated facilitating bladder neck reconstruction, then reassessing again after urology team start and pubic diastasis is still eliminated after dissection.

Patient evaluation

1. History: complete of any other medical or surgical condition
2. Exam: complete general examination, followed by local exam of the diastasis
3. Radiology: Plain radiograph of the whole pelvis AP to measure pre-operative inter-pubic -closest interpubic distance- and inter-ischial distance at the same level of which inter-pubic distance was measured to calculate IS/IP ratio

Peri-operative protocol

All operations were done with single team pediatric orthopedics and single team pediatric urology, during period October 2020 till February 2024.

Pre-operatively clinical reassessment of patients was carried out regarding pubic diastasis clinical value

Intra-operative

1. In supine position patients underwent formal Sponseller's osteotomy (Gearhart et al., 1996)
2. followed by urology team intervention
3. application of the spica cast which is usually done in the position of flexion, slight abduction 10° and internal rotation, the application of 3-4 layers of plaster of Paris followed by fiberglass 1-2 layers, using bar connecting both lower limbs positioned in front of flexed knees, then making V shaped opening starting at lower most abdominal part of the cast with limbs extending cranially but ensuring to spare ASIS coverage, the cast extended from below costal margin proximally till above malleoli distally sparing ankles (Figure 3).



Figure 1: clinical interpubic distance



Figure 2: Markings over FA in blue ASIS and planned skin incision in black

Post-operatively: immediate post-operatively to assure cast instructions of letting the heel free by slight raising legs

1. follow up was carried out on 2nd post-operative day clinically to check on the urology wound, then again in 1 week.
2. then every 2 weeks 1st to assess clinically urological wound closure integrity
3. 2nd on 4th week post-operatively to radiologically assess osteotomy union and to remove cast if complete union is achieved usually this visit is done for children aged up to 1 year
4. If complete union wasn't obtained in 4th weeks visit another visit after 2 weeks (on 6th week) to remove the cast after radiological evaluation
5. then regular follow up every 3 months twice then every 6 months up to 2 years post-operative.

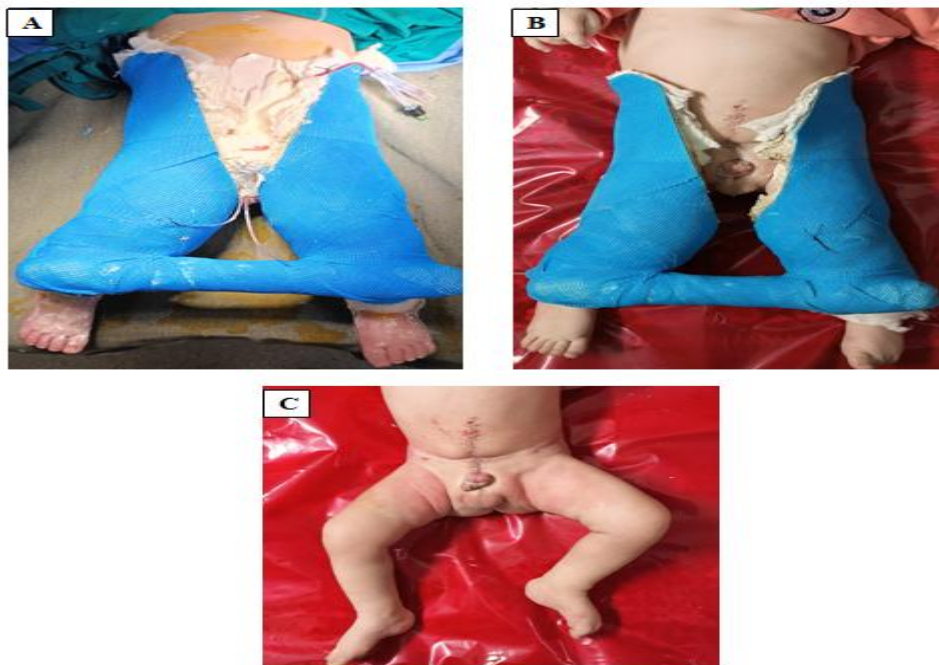


Figure 3: Immediate post-operative photo of the newly designed cast A, at the end of 4 weeks follow up and complete successful healing of urological wound and osteotomy site B, after cast removal showing successful closure, healing and no skin breakdown

Regarding spica cast, it offered better mother to child bond regarding carrying the child and moving him much more freely than with the aid of external fixator especially during feeding children below age of 2 who still in need of breast feeding, spica cast offered easily access and carrying instead of using bottle or food to those patients near weaning age.

Cast shape and configuration evolved throughout the whole period to meet our needs for appropriate care to the urological wound and avoiding failure and cast complications from soaking and one of the most devastating complications; Cast syndrome which is resembling intestinal obstruction in presentation and management (figure 4)

Cast syndrome is duodenal 3rd part obstruction between the acute angle made by superior mesenteric artery and abdominal Aorta by a nut-cracker effect, it's also called superior mesenteric artery syndrome -SMA syndrome- was first discovered in scoliosis patients who underwent Harrington rod instrumentation and plaster of Paris body plaster jacket application, it was managed conservatively with a nasogastric tube, intra-venous fluids and NPO for 3 days.

The final cast shape had 0% rate of cast syndrome and of cast failure and break down.



Figure 4: Cast Evolution showing A; the early spica cast following formal design, B: the earliest case put in cast with the complete anterior opening, C final shape of the new design



Figure 5: Journey of 3 years old boy pre-operative A, then post-operative in new -KZ- cast B, then after complete wound healing, bony union and cast removal after 6 weeks C

Assessment:

Success of pelvic osteotomy-assisted closure of exstrophy is defined as closure of urological wound with insertion of the bladder into abdominal cavity without development of: wound dehiscence, prolapse or vesico-cutaneous fistula requiring re-operation with re-osteotomy.

Failure was defined as urological wound dehiscence due to failure of achieving tension free repair and closure in the form of bladder dehiscence, prolapse, vesico-cutaneous fistula requiring re-operation with re-osteotomy.

Delayed or 2ry exstrophy or bladder extrusion again from abdominal cavity wasn't mentioned in literature so urological wound healing meant success of the osteotomy to achieve its goal.

Regarding 1ry outcome all cases achieved fully osteotomy union at 4-6 weeks post-operatively with 0 percent non-union and delayed union rate.

About 2ry outcome none of the patients had:

SMA syndrome, cast breakdown or need to be changed, femoral nerve palsy on assessment after cast removal, sciatic nerve palsy, pressure ulcers, or compartment syndrome.

Some of patients had minimal skin rash on cast removal which responded to topical treatment for maximum 3 days, 1 patient had superficial fungal infection of skin due to poor hygiene but it responded to topical treatment and resolved within 1 week.

RESULTS

We conducted this prospective retrospective study on bladder exstrophy patients from the year October 2020 to February 2024, in a specialized pediatric orthopedic unit in collaboration with a pediatric urology unit at a North African tertiary university hospital -Mansoura university, Egypt-by single team pediatric orthopedics and single team pediatric urology -each team had one consultant with experience of more than 10 years in such field and 1 specialist-, the study included 18 patients who underwent 21 osteotomy assisted closure of exstrophy, 7 girls

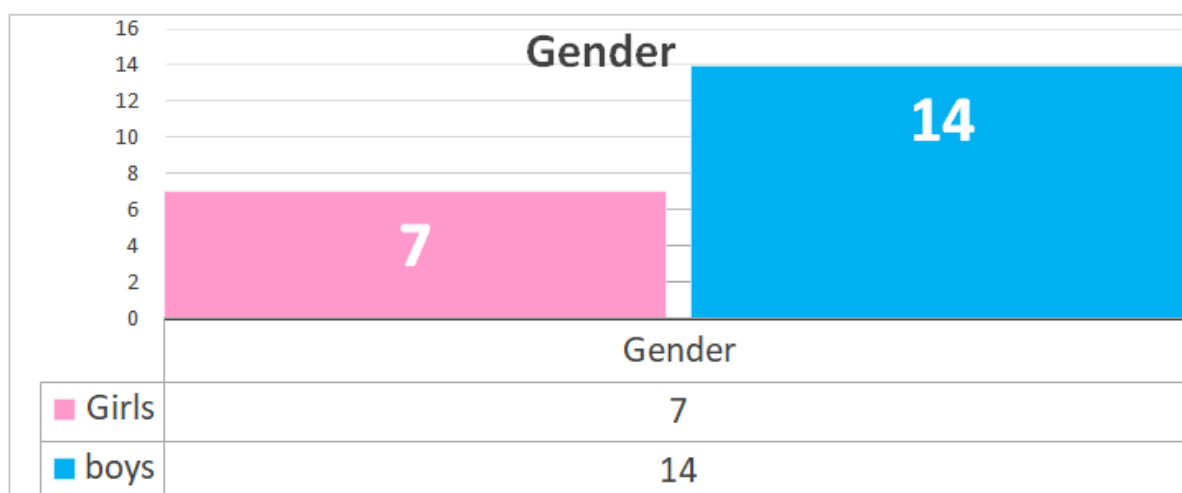


Chart 1: Gender distribution

33.33% and 14 boy 66.67% (chart 1), age of patients at time of intervention varied from 3months to the age of 25 months.

Statistically analyzed and also correlated to clinical opinion of the urological team of the ease of pelvic manipulation and closure intra-operatively.

Follow up period from 2 days and 14 days post-operatively ensuring complete closure of urological wound and excluding any wound dehiscence which necessitate orthopedic re-intervention up to 3 years in some cases, it wasn't a routine to follow up patients for longer period as delayed wound dehiscence didn't occur in our series or in the literature review, yet due to frequent urological follow-ups we followed up cases up to 7 years.

Success of pelvic osteotomy-assisted closure of exstrophy was defined as closure of urological wound with insertion of the bladder into abdominal cavity without development of: wound dehiscence, prolapse or vesico-cutaneous fistula requiring re-operation with re-osteotomy.

While spica cast success is to achieve bony healing in appropriate time of 4-6 weeks post-osteotomy without delayed or non-union at osteotomy site, Success rate of spica cast to achieve bony healing was 100%, while success rate of osteotomy assisted closure in the 21 closures was 90.48%.

None of the patients had:

SMA syndrome, cast breakdown or need to be changed, femoral nerve palsy on assessment after cast removal, sciatic nerve palsy, pressure ulcers, or compartment syndrome.

While Some of patients had:

minimal skin rash on cast removal which responded to topical treatment for maximum 3 days, 1 patient had superficial fungal infection of skin due to poor hygiene but it responded to topical treatment and resolved within 1 week.

DISCUSSION

Pelvic osteotomies has been adopted aiding closure of bladder exstrophy as old as 1892 when first was done by Trendelenburg, also following pelvic osteotomies, he made a device called it "pelvic sling" to immobilize osteotomy and approximate pubic bones, yet his 1st patient passed away because of post-operative anemia but the idea since then was implemented by many surgeons, and till the present day is considered the cornerstone of giving the congenitally externally rotated pelvis and deficient anterior abdominal wall the malleability required for successful tension free closure. (Trendelenburg, 1892, Haffar et al., 2023a)

Studies showed that type of osteotomy and means of immobilization have no impact on success of closure in the neonatal population -below 28 days old-, and in the same study it showed that the old posterior osteotomy although being historical, demanding but had favorable outcome regarding closure success, but lengthy operative timing requiring prone position first then repositioning into supine intra-operative, the oblique iliac osteotomy adopted by some European centers showed favorable outcome regarding closure success, the combined osteotomy offers best results in all age groups with ease of access of the patient being supine for both orthopedic and urological teams, also give an access to anterior osteotomy and the post para-sacroiliac vertical limb osteotomy offering more malleability of the pelvic ring for internal rotation and anterior approximation which is the same finding in our current study supporting the combined osteotomy. (Khandge et al., 2021)

Although anatomic complete reduction of inter-pubic distance and obtaining the normal anatomical apposed symphysis intra-operatively is mandatory for urological reconstruction, which is the function of the osteotomy and then to obtain tension free closure for its success, here maintaining this normal inter-symphyseal distance till closure success is the function of means of immobilization in the post-operative period. (Haney et al., 2023)

Hip spica, Bryant's traction, modified Buck's traction, mummy wrap and external fixator all are methods were described used and studied thoroughly in literature, with pros and cons for each regarding lengthy hospital stay and femoral nerve palsy, pin tract infection and difficult manipulation and movement regarding external fixator group associated with traction, mummy wrap offers too limited immobilization with less pain control, spica cast complications include skin rash or break down, cast syndrome -SMA syndrome-. (Shnorhavorian et al., 2010, Khandge et al., 2021, Wild et al., 2011)

Despite of long hospital stay associated with traction post-operatively -4-6 weeks- and necessity of hospital admission to offer facilities for traction, but modified Buck's traction showed superior results in maintaining the intra-operative position of the osteotomized bone, no skin break down and being superior to spica or mummy's rap according to recent 2023 study on over 700 cases, while other studies showed no difference between those mentioned methods of post-operative osteotomy. (Haffar et al., 2023b, Sack et al., 2015, Arlen et al., 2011)

Regarding means of immobilization in cases underwent immobilization with spica cast had better psychological state noticed with less crying and irritability most noticed with ease of mother carrying them, which was also noticed in other studies and termed bonding with family and the mother. (Haffar et al., 2023a, Zaman et al., 2019)

Also cost of treatment is an important point of interest of which in our institute, long term follow up of older cases we concluded that the burden of repeated surgeries and disabilities of prolonged follow up and repeated procedures required for cases failed are not comparable to the cost of adding orthopedic pelvic osteotomy step for urological reconstruction and ease of tension free closure in a complete primary repair procedure or even staged modern staged repair which was the same outcome in other studies, side by side with spica cast being easiest means of immobilization for parents handling and least hospital time needed before discharging the patient. (Haffar et al., 2023a, Hesh et al., 2016)

CONCLUSION

The newly designed spica cast of exstrophy -KZ cast- is a safe effective means for post-operative immobilization following exstrophy closure with less complications than old design.

Informed consent

All patients' parents or care-givers signed an informed consent after complete explanation of the procedure with its possible complications.

Conflict of Interest

All of the authors declared no conflict of interest.

Financial Disclosure

The authors declared that there was no financial support in this study.

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