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BLADDER EXSTROPHY REPAIR

Assiut Experience.

CONCEPT, PRACTISE AND FACT
Bladder extrophy exists as part of a spectrum of anomalies which includes cloacal extrophy at one extreme and glandular epispadias at the other.

First description of the bladder extrophy appears on an Assyrian tablet from 2000 BC preserved in British museum in London.
* In all cases of reparative surgery, in which the defect is congenital, aim of the surgeon should be to restore the parts to their normal relations as nearly as possible.

* Many techniques and their modifications were described in a trial to achieve those goals.

* The best operation would be one that reverses the condition of the accident.

{Cantwell.1895}
*Goals of reconstructive surgery of bladder exstrophy are well established, however to our knowledge no ideal surgical reconstructive technique was settled to reach those goals.

*Staged repair of bladder exstrophy reported a satisfactory outcome in a large series of cases with long term follow up, in the mean while recent reports of one stage repair were encouraging.
Ideal management of bladder exstrophy?

• Early cystectomy and urinary diversion

• Functional reconstruction

YES ✓ ✓ ✓
The management of the bladder extrophy has evolved from early cystectomy and urinary diversion to the functional closure of bladder, that remains the golden standard methodology for achieving the aim of appropriate bladder closure.

(Gearhart, 1999)
CONCEPT OF REPAIR

I-Secure abdominal wall closure.
II-Urinary continence (volitional voiding) with preservation of renal function.
Concept of repair

Functionally and cosmetically accepted external genitalia.
CONCEPT OF STAGED REPAIR

- Proper functional bladder closure is the cornerstone for successful repair.
- Bladder exstrophy at birth has maturational delay and so sensitive for outlet obstruction.
- Primary defect of bladder exstrophy is multifactorial and not just a herniation.
CONCEPT OF SINGLE STAGE

*Primary defect of bladder exstrophy is anterior abdominal wall herniation, thus the bladder and urethra must be treated as single unit.

*Early reconstruction would allow for mechanical cycling of the bladder in 1st year of life, that enhance restoration of urinary continence.

*Complete repair could be achieved in single stage.
PRACTISE

Nothing new under the sun
Exstrophy repair

Staged

1 stage
**Schools** OF Bladder extrophy repair:

I-Staged : *(Jeffs et al. 1972)*

- Primary bladder closure
- B.N reconstruction.
- Epispadias repair.

I-One stage.
Schools of extrophy repair:

I- Staged repair:

1a: Johns Hopkins approach (Gearhart et al. 1989)

- Bladder closure at birth.
- Epispadias at 6–12 months.
- BN repair at 4-5 ys.

1b: Warsaw approach (Baka-Jakubiak et al. 2000)

- Bladder closure at birth.
- Epispadias & BN repair at 4-5 ys.
Schools of extrophy repair:

II-One stage repair:

- **1a**: Seattle approach *(Mitchell et al. 1999)*
  
  Bladder closure, Epispadias repair (without BN repair) at newborn.

- **1b**: Erlangen approach *(Rosch et al. 2001)*
  
  Bladder closure, epispadias & BN repair with ureteral reimplantation at infancy.
Q- With or without osteotomy?

- Time of surgical reconstruction. ✓
- Extent of pubic diastasis. ✓
- Malleability of the pelvis?
- Size of bladder template?
Advantages of Osteotomy:

- Diminishes the tension on abdominal wall closure → # dehiscence.
- Placement of posterior urethra and bladder neck deep within pelvic ring → continence.
- Bringing of pelvic floor musculature near the midline to support the bladder neck → continence.
- Hip force ↓ - stress & degenerative
Types of osteotomy:

A- Posterior iliac osteotomy.
B- Anterior transverse innominate osteotomy
   Better approximation.
   No turn of patient.
C- Combined anterior osteotomy.
D- Others.
One stage bladder exstrophy repair:

• Not new:!!!

- Trendlenberg, 1906
- Young 1942
- Schultz 1958
- Ansell 1971, 1979
- Montagnani 1982
- Gearhart 1991
- Fuchs et al, 1996
One stage bladder exstrophy repair:

Results of single stage repair:

*Continence: 0-45% (17%)

*Renal damage: 90%

**Grady and Mitchell, 1999

* Continence: 80-90% dry
  with no renal damage
One stage bladder exstrophy repair:

- Closure of bladder plate:
  incontinuity with bladder neck and urethra (with no antireflux procedure?)

-EpisPADias repair:
  Mitchell technique of disassembly
  Modified Cantwell-Ransley repair
Surgical Steps:

I-Bladder dissection:

Ant.  Lateral  Posterior
 Radical dissection of the bladder
Prognostic factors to achieve continence:

* Early closure (Urodynamic features, Borer et al., 2005)

* Most significant factor is bladder capacity (85cc & 115cc)

* Role of osteotomy (MR for pelvic floor, Gargollo et al., 2005)
Surgical Steps:
II - Epispadias repair.

- Complete penile disassembly (CPD)
- Modified Cantwell-Ransley
The two corporeal bodies are supplied by deep penile arteries.

Additional pudental branches, the dorsal arteries of the paired neurovascular bundles supply the glans penis.

Therefore, it is possible to disassemble the epispadiac penis.
Mitchell and Bagli (1996): described their technique of complete penile disassembly for epispadias repair.

Original (CPD)

- 1998: Zaontz et al. JU 17 boys
- 2001: Caione et al. JU 17 boys
- 2003: Hammouda. JU 42 boys
Complete penile disassembly (CPD)

- Penile dissection from ventral aspect.
- Preservation of skin attachment of plate.
- Directly on Buck's fascia overlying urethral wedge.
- Medial side directly on tunica albuginea overlying each corpus.
- Plate is tubularized using interrupted sutures.
- Secured to tip of glans and meatoplasty is completed.
Modifications of the original technique were described:


EDITORIAL COMMENT

Bladder extrophy remains an uncommon and severe genitourinary anomaly. These authors present the largest series of patients to date who have undergone extrophy reconstruction using the complete primary repair technique developed by Mitchell and Bagli as a concept evolution of the penile disassembly technique created to reconstruct the epispadic penis (reference 3 in article). The authors consistently applied the principles of 1) complete bladder, bladder neck and urethral mobilization, 2) deep pelvic dissection and 3) anatomical positioning of the bladder and urethra in 33 children.

During a median 4-year followup more than 70% of the children in this series achieved some degree of urinary continence, including some without the need for bladder neck reconstruction. Few of these patients had complications and no patient experienced loss of a corporeal body or glans. Surprisingly few children in this series had hydroureteronephrosis. As importantly, less than 10% of the children reported on required bladder augmentation to become continent, in contrast to other series with reported rates of augmentation of almost 30% to become dry.

These results demonstrate the potential of this surgical technique when applied consistently by surgeons well versed in the principles of the complete primary repair technique. The authors present excellent results in this large series.

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Hammouda and Kotb reported a series of 33 cases. They achieved continence in 72% of patients in their series with minimal complications, mirroring results at our institution. Hammouda has also used the complete penile disassembly technique in 42 patients with excellent functional results. Borer et al also reported this.
Why there is a debate?

* Complex procedure
* Reflux (10-50%)
* Experience (pediatric team)
* Premature babies & kids with small penis
Why there is a debate:

I-Bladder dissection:

*Technique:
- Same rules
- One stage # Modern version of staged
- waiting strategy ????
Continence after exstrophy repair:

<table>
<thead>
<tr>
<th></th>
<th>Continence</th>
<th>Renal damage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Staged repair</td>
<td>88% (10-30%)</td>
<td>13-20</td>
</tr>
<tr>
<td>One stage repair</td>
<td>65.5%-80%</td>
<td>0</td>
</tr>
</tbody>
</table>
Why there is a debate:

II-Epispadias repair:

*Complete disassembly

#Modified Cantwell-Ransley (Incomplete disassembly)

*Complete disassembly:

- Low fistula rate
- Incidence of hypospadias (avoided)
- Ischemic changes ?? (avoided)

**LEARNING CURVE**
CPD

- Correct plane.
- Interrupted fine urethral sutures.
- Fixation sutures.
- Urethral mesentery.
- Shift.
- Not in small sized and premature babies.
- Modified Cantwell Ransely.
- CPD is not without worries.

- CPS is an effective alternative to MCR.
-Neither Mitchell or Modified Cantwell Ransely can normalise the penis.

(Mouriquand, 2003)
Should I change the technique I already use and give good results?
FACTS

- Repair of bladder exstrophy is not for occasional surgeon.
- More than one operation may be needed.
- Rules of bladder dissection are established.
- Bladder has maturation delay and sensitive to outlet obstruction, however cycling in 1st year is important.
- Osteotomy has advantages.
- One stage repair is not new, has its indications.
FACCTS

- BNR is complementary in some cases.
- CPD is not without worries, slightly differ from MCR (0.5 cm), but has lowest fistula rate.
- Dry interval is not true continence.
- Dry interval term (social continence) has fallacies.
- Learning curve, pain free and good nursing during postoperative period are essentials.
FACTS

- BNR is mandatory in some cases.
- CPD is not without worries, slightly differ from MCR(0.5 cm), and has lowest fistula rate.
- Dry interval is not true continence.
- Dry interval term (social continence) has fallacies.
- Parameters for success should be addressed.
- Learning curve, pain free and good nursing during postoperative period are essentials.
FACTS

- Young-Dees-Lead better technique and its modifications are preferable in bladder exstrophy complex, aiming at volitional voiding.

- Continent catheterizable stomas are valid practical treatment, while injection therapy are good promising options.
Requirements:

- Pediatric Urologist
- Anaesthesia
- Nursing

Learning curve
THANKS