PD-112

CHALLENGES IN MANAGEMENT OF CAUDAL DUPLICATION SYNDROME

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INTRODUCTION

- Caudal Duplication Syndrome (CDS) is a rare and complex anomaly.
- Incidence < 1 in 100,000 cases
- ~ 100 cases reported.
- Term CDS was proposed by Dominiguez et al. in 1993 (though similar manifestations rep. historically since 18th century)
- An association between gastrointestinal, genitourinary, and distal neural tube malformations.
- Recently - Associated musculoskeletal & cardiovascular
- Each case is unique and adds knowledge to the available literature

METHOD

A Prospective case series of CDS is described.

Clinical Features

Case 1

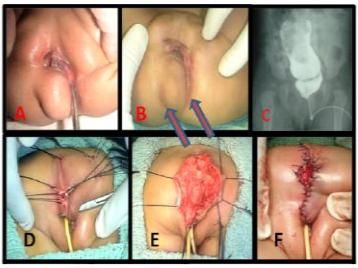
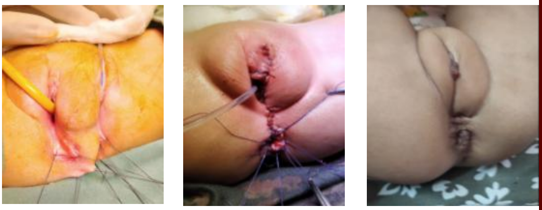


Figure1. A: Perineallipoma with Hegar dilator in perineal fistula. B: Split

Case 2 - cloacal duplication



Complete repair with removal of the intervening lipomatous mass and approximation of the divaricated anal sphincters on either side

Case 3

Colostomy -> Complete repair with removal of the intervening lipomatous mass and approximation of the divaricated anal sphincters on either side. He expired after surgery due to respiratory failure with seizures.

Case 4

Boy underwent excision of a prolapsing mass arising from between the two scrotal sacs that was a benign teratoma on histology.

RESULTS

- All had associated ARM. 2 were females.
- Colostomy done in 3.
- Both girls underwent complete repair with removal of the intervening lipomatous mass and approximation of the divaricated anal sphincters on either side. Postoperatively, both children were continent and could contract their rectum.

Case	Gender	Lower Gastro-Intestinal tract	Genitourinary Tract	Spinal Anomalies	Intervening perineal mass	Other associated anomalies
1	Female	ARM	Duplicated Vagina, vaginal septum	Dermal sinus, spina bifida, intraspinal lipoma	Lipoma	
2	Female	ARM with Cloacal malformation Duplicated rectum	Duplicated vagina	Filum lipoma	Lipoma	
3	Male	ARM wih recto-bladder neck fistula	Diphallia Scrotal hypospadias Penoscrotal Transposition Bifid scrotum	Not evaluated	Hamartoma- Firm cartilaginous, mucosa lined	Cleft palate, TMJ ankylosis
4	Male	ARM (under investigation)	Diphallia Scrotal Hypospadias Penoscrotal Transposition Bifid scrotum	Not evaluated	Teratoma	

	Age	Treatment	Colostomy	Follow Up
Case 1	8 months	PSARVP and excision of mass	No colostomy	Continent at last 13 years follow up (till 2022)
	48 months	Anoplasty for anal stenosis leading to rectal ectasia		
Case 2	Newborn	Colostomy	Yes	Continent at 6.5 years age (6 years follow up)
	11 months	ASARVUP and excision of mass		
	1 year 10 months	Colostomy closure		
Case 3	Newborn	Colostomy	Yes	Difficult intubation due TMJ Ankylosis
	18 months	AP PSARP and excision of mass		Operated In supine as not possible to make prone due to threatened airway Expired after surgery due to respiratory failure with seizures
Case 4	Day 2	Transverse Colostomy	Yes	Awaiting PSARP
	Day 9	Excision of perineal mass		

CONCLUSION

- CDS is a complex malformation requiring a planned treatment.
- The anal sphincter complex should be identified carefully during surgery and placed around the neoanus for the child to attain continence. Associated anomalies may account for mortality.