

PROTOCOLISED AND MULTIDISCIPLINARY TEAM APPROACH IN THE MANAGEMENT OF CLOACAL MALFORMATIONS – EARLY EXPERIENCE

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Aim: We present our outcomes of children with cloaca managed through a protocolised pathway by a dedicated multidisciplinary team (MDT).

Methods: A protocol based management was instituted from birth. The MDT comprised of Paediatric Surgeon and Urologist, Nurse specialists (Urodynamics & Stoma), Urodynamics expert, Psychologist and Nephrologist. Anatomical & functional classification was adapted to prospectively categorize patients, monitor the results and enable comparison of outcomes (Table 1).

Results: (Table 1) Number of patients treated in 4 years(2012-2016) n=16. 14 had reconstruction. 2 await reconstruction. Mean age at primary reconstruction was 11 months (Range 4-26). Three were unsuitable for perineal urethral channel. Two(IL, III) with poor bladder neck and pelvic floor musculature had bladder neck closed with formation of appendico-vesicostomy; 1(III) with bladder agenesis had an ileal conduit.

Anatomical outcome: Of the remaining 11[6(IS),4(IL),1(II)] normal perineal anatomy was restored in 10. 1(IL) developed urethral stricture requiring subsequent appendico-vesicostomy.

Bladder function: 9/11(73%) [IS(5), IL(3), II(1)] void spontaneously with good emptying and no UTI. 1/11(9%) developed urethral stricture (IL) and one incomplete bladder emptying (IS) with recurrent UTI. 2(IL, III) are on CIC via appendico-vesicostomy and 1(III) has an ileal conduit.

Bowel function: 4/14 (28%) opening bowels spontaneously. 3 (21%) maintained satisfactorily on enema program. 3 (21%) dependent on combination of laxatives and enema. Anal stenosis 1(9%) and mucosal prolapse 1(9%). Two await colostomy closure.

Renal function: 14/16 have normal renal function which is monitored regularly. 2of16 (CKD1, CKD3) have underlying renal dysplasia.

Conclusion: The early renal outcomes managed through the MDT are encouraging. Further follow up and comparative data is awaited. The variation in anatomy is complex and the value of an experienced MDT, protocolised management and categorisation is crucial to maximise good long term functional outcome and have comparative data. We recommend the categorisation we have utilised to facilitate this.

Table 1: Anatomical & Functional Classification, Associated anomalies and Primary operation.

Type	Category Code	n=16 n(%)	Associated anomalies – n(%)					Primary reconstruction 14 (2 WL)
			Spine (9)56	Mullerian (12)75	Cardiac (10)62	Renal (9)56.2	Misc	
Type I Classic Cloaca	IU	-	-	-	-	-	-	-
	IS	8(50)	5 (ISS)	5(1 MA, 2MH,2MD)	5	4	OA	3 PUM 3 TUM
	IL	5(32)	3 (ILS)	4 (1MH, 1MD,2MD+ MH)	3	3	OA	4 TUM 1 UC
Type II Posterior Cloaca	IIC (Classic)	II	-	-	-	-	-	-
	IIV (Variant)	IIV 1(6)	-	1(MA)	1	-	-	1 TUM
Type III Uncategorised	III	2(12)	1 (IIIS)	2 (1MA,1MD)	1	2	-	2 UC

Legends:

CC – Common Channel, IU – Ultrashort CC (<1cm), IS – Short CC (1-3cm) ; IL – Long CC (>3cm), ISS – Short CC with Spinal involvement, ILS – Long CC with spinal involvement.

V- Variant, MA- Vaginal atresia, MH- Hydrocolpos, MD – Mullerian duplication,

WL – Waiting list, OA – Oesophageal atresia

PUM – Partial Urogenital Mobilisation with PSARP, TUM – Total Urogenital Mobilisation with PSARP, UC – Unclassified.