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#### A condition is for life..... not just paediatrics

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> University College Hospitals NHS Trust NHS NHS Foundation Trust



### Historical phases

### **L**UCL



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# Forward thinking.....



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### **CONGENITAL LIFELONG UROLOGY**





### **CONGENITAL LIFELONG UROLOGY**

### **WORKING GROUP**





### CLU objectives



To form and work as a multidisciplinary group aimed at improving the care of patients with congenital urological anomalies.

To attract interested practitioners into training in and taking on this work as part of their remit.

To form and formalize a network of interested practitioners with the potential for clinical networking that may include (for example) electronic MDT meetings for complex case discussion

The potential for development of treatment guidelines/standardization and metrics.

The development of education in the care of these patients – this may be achieved through fellowship programmes, other opportunities such as course may also develop.



### CLU Objectives



**Multidisciplinary** 

Get more people

**Network** 

Guide and provide standards



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### Unspoken (un)truths:

Peds	Adult
I will always be here for you	
Nobody does it better	
Of course, we can talk about sex	
The adult team will fix that	
Any problems just call	
	What the f*** do I do with that?
	Yes, yes there will be space



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### Unspoken (un)truths:

Peds	Adult
I will always be here for you	He retired?????
Nobody does it better	Nobody does it
Of course, we can talk about sex	Now, I do know something about that
The adult team will fix that	They said what?
Any problems just call	Who?
We could help?	What the f*** do I do with that?
	Yes, yes there will be space





### WHY IS THIS IMPORTANT? – some examples Like how many get lost Different priorities Get rid of families





### WHY IS THIS IMPORTANT? – some examples How many get lost?

25-50 % if no good tx clinic Falls to 7.9 % with Tx Clinic Gaydos et al 2020 Relationships Sex

Privacy

Shifting control but not removing support





# Transition







### Four Models



- 1. Paeds urologist follows the patient through in an integrated department
- 2. Paeds urologist continues to follow their patient into a separate adult department
- The patient moves in adolescence to see an adult specialist in congenital urological disorders
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- 4. The patient sees adult practioners when needed







# Teaching the young adult how to be independent





National Service Framework for Children, Young People and Maternity Services

Every Child Matters - Change for Children

#### Transition: getting it right for young people

Improving the transition of young people with long term conditions from children's to adult health services Adult Health Se Employment Further Educat Children's Heal School



Adolescent transition care

Guidance for nursing staff

department for children, schools and families



#### Transition: moving on well

A good practice guide for health professionals and their partners on transition planning for young people with complex health needs or a disability

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#### Ready Steady Go: moving through the programme.

#### **Ready Steady Go: Moving through the programme** Ready Steady Go: Each Young person (YP) progresses at their own pace



Arvind Nagra et al. Arch Dis Child Educ Pract Ed 2015;100:313-320

EP

# Improving the transition between paediatric and adult healthcare: a systematic review

R Crowley, I Wolfe, K Lock, M McKee

Arch Dis Child 2011;96:548-553. doi:10.1136/adc.2010.202473

Systematic review
Identifies 10 studies
6 studies show significant improvement
Conclusion: strategies of succesful programmes Include patient education and transition clinics.

JCID

What is already known on this topic

- The transition to adult services can result in poor health indicators and loss to follow-up for adolescents with chronic disease.
- Transition is now recognised as a key component of care, across the spectrum of physical and mental illness and disability, though there has been little high quality evaluation published.

#### What this study adds

- Most studies evaluating transitional care have been for patients with diabetes mellitus, with programmes targeted at improving patient education, staff continuity or service delivery.
- Existing evidence supports the use of educational programmes, joint paediatric/adult clinics and specific young adult clinics.



### Background reading

### **L**UC

### J Urol. 2012 Apr;187(4):1164-71. doi: 10.1016/j.juro.2011.12.011. Epub 2012 Feb 14. Adult care of children from pediatric urology. Woodhouse CR<sup>1</sup>, Neild GH, Yu RN, Bauer S.





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### What have we learned?

### **UCL**





### On the shoulders of giants

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### Survey of need??? BAPU 2013 Wood et al Unpublished



33 Paediatric urologists\*\* – 100% response rate
63 % refer at adolescence (18 % continue care)
Appropriate age for referral 12-21 yrs
39 % refer to adult
36 % to adolescent
61 % have formal adolescent MDT
58 % said there was a local transition service

Adolescent care best by adolescent urologists (Preference adolescent>paeds>adult)

\*\*most from specialist centres







### WHY BOTHER?





CIP

#### What do we see?



With thanks **Peter Cuckow Stuart O'Toole Christopher Woodhouse Chris Kimber** 

Malignancy<sup>23</sup>







**Differences of** sex development







Complications



Incontinence



Exstrophy





# The importance of multidisciplinary care





### What do you tell the parents?

### **L**









#### Presentations have changed

### **LICL**





### Presentations have changed

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### ?Outcomes..... have not



FIG. 4. Redrawn from [1]. Age at follow-up of 88 patients who survived beyond the age of 12 years and in whom the outcome is known.







### **NEUROPATHIC BLADDER**

### Which aspects are lifelong?

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### Which aspects are lifelong?







### What do we learn?





### **Consider executive function**





#### Limitations in

- Intellectual function
- Initiation of tasks
- Problem solving
- Decision making
- Emotional regulation



#### Augmentation Cystoplasty and Risk of Neoplasia: Fact, Fiction and Controversy

#### T. T. Higuchi, C. F. Granberg, J. A. Fox and D. A. Husmann\*

From the Department of Urology, Mayo Clinic, Rochester, Minnesota

0022-5347/10/1846-2492/0 THE JOURNAL OF UROLOGY® © 2010 by American Urological Association Education and Research, Inc. Vol. 184, 2492-2497, December 2010 Printed in U.S.A. DOI:10.1016/j.juro.2010.08.038



Table 1. Characteristics of patients with congenital bladder anomalies and bladder cancer

Pt No.	Etiology of Bladder Dysfunction	Age Diagnosis (yrs)	Yrs From Augment to Ca*	Augmenting Segment	Bladder Calculi History	Ca Type (TNM stage)	AJCC Stage	Survival
			F	ts with augmenta	ation cystoplasty			
1	Neurogenic bladder	61†	52	lleum	No	Urothelial (pT2, N0, M0)	2	NED 72 mos
2	Neurogenic bladder	64†	53	lleum	No	Urothelial (pT2, N0, M0)	2	NED 84 mos
3	Posterior urethral valves‡	43	25	lleum	No	Adenoca (pT3, N2, M0)	4	Died of disease 12 mos
4	Posterior urethral valves‡	34	22	Colon	No	Adenoca (pT3, N1, M0)	4	Died of disease 24 mos
5	Exstrophy	51	32	Colon	No	Adenoca (pT3, N2, M0)	4	Died of disease 36 mos
6	Exstrophy	52†	47	lleum	Yes	Adenoca (pT3, N2, M0)	4	Died of disease 18 mos
7	Exstrophy	30	22	lleum	No	Adenoca (pT4, N2, M1)	4	Died of disease 6 mos
			Con	trols on intermitt	tent catheterizatio	n		
1	Neurogenic bladder	62	Not applicable	Not applicable	Yes	Squamous cell (pT3, N0, M0)	3	Died of disease 24 mos
2	Neurogenic bladder	38	Not applicable	Not applicable	No	Urothelial (pT4, N1, M1)	4	Died of disease 10 mos
3	Neurogenic bladder	55	Not applicable	Not applicable	No	Urothelial (pT3, N1, M0)	4	Died of disease 24 mos
4	Exstrophy	44	Not applicable	Not applicable	No	Adenoca (pT3, N2, M1)	4	Died of disease 8 mos

\* Interval from initial ileal conduit formation to diagnosis of cancer.

† History of ileal conduit with subsequent urinary undiversion with incorporation of conduit into bladder.

‡ History of renal transplant on immunosuppression.



#### Renal status Malakounides et al 2013

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Table 1 R	Renal function Ref. [19].	
CKD stages	Description (corrected GFR in ml/min/1.73 m <sup>2</sup> )	%No. of patients (total 120)
Normal	Normal GFR and imaging $(\geq 90)$	48% (58)
I	Kidney damage with normal GFR ( $\geq$ 90)	6% (7)
II	Kidney damage with mild reduction of GFR (60-89)	36% (43)
Ш	Moderate reduction of GFR (30-59)	8% (10)
IV	Severe reduction of GFR (15-29)	0.8% (1)
V	Kidney failure (<15 or dialysis)	0.8% (1)

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#### Cause of death Malakounides et al 2013

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Cause of death	Age
Sepsis (hindquarter amputation)	14
Sepis (post ileal conduit)	17
Sepsis (pressure sores)	??
Sepsis (pressure sores)	??
Blocked shunt	11
Not given	
Not given	

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Mean age of death 14 (0-25 years) 4.4 % NONE FROM RENAL FAILURE

Previous series upto 1/3 death from renal failure (Singhal et al 1999)



### Familial Risk

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Relationship	Incidence per 1000 LB
General Population	0.7-1
Mother + 1 affected child	20-50
Mother + 2 affected children	100
Patient with myelodysplasia	40
Mother over 35 yrs	30
Sister of mother with affected child	10
Sister of father with affected child	3
Nephew who is affected	2

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#### FOLIC ACID Affected family 4mg OD for at least 2 months pre conception General Pop' n 400 µg OD Reduction by 50%





### Hypospadias



### Testosterone makes no difference



Testostrone makes no difference To penile length or outcome

Proximal hypospadias is **negatively** associated with penile length



### Length of follow-up is key

# LICL

Study	Pts	Meatal location	Repair type	Complication n (%)	Follow-up (mo)	Fistula	Dehiscence	Meatal stenosis	Stricture	Diverticulae	Residual chordee
Snodgrass, 2011 [41]	26	Proximal	TIP	3 (13%)	2 38	0	2	0	1	0	0
Ghanem 2010 [42]	49	Proximal	TIP	5 (12%)	16-72	4	1	1	0	0	0
Cheng 2002 [2]	100	Midshaft/ proximal	TIP	4 (4%)	4-66	3	0	1	0	0	0
Braga 2007 [43]	75	Proximal	TIP 35	TIP 21 (60%)	6-80	15	3	1	0	0	2
			Onlay 40	Onlay 18 (45%)		8	2	1	2	0	5
Ghali 1999 [44]	148	Midshaft/ proximal	Tubularized preputial flap	48 (32%)	6-53	22	?	17	13	7	3
De Mattos e Silva 2009 [45]	126	Proximal	Onlay	34 (27%)	1-97	18	13	0	2	1	?
Stanasel 2015 [26]	56	Proximal	Two-stage flap	38 (68%)	38.6 (mean)	32	2	5	8	8	?
McNamara 2015 [25]	134	Proximal	Two-stage flap	71 (53%)	1-260	39	19	17	16	12	3
Zheng 2015 [46]	66	Proximal	One-stage	8 (25%)	20-60	5	1	0	6	0	?
			Two-stage	8 (25.5%)	18-60	7	1	1	0	0	?
Long 2017 [47]	167	Proximal	One-stage	53 (62%)	13-71	40	4	13	0	8	6
			Two-stage	40 (49%)	14-44	30	6	5	0	5	3
Salle 2015 [27]	140	Proximal	TIP	35 (61%)	6-123	18	7	6	3	1	8
			Inlay graft	12 (52%)	8-91	3	4	3	1	0	4
			Two-stage	23 (38%)	6-127	7	10	0	1	1	3
Ferro 2002 [48]	34	Proximal	Two-stage graft	8 (24%)	1-48	2	4	0	1	1	0
Johal, 2006 [49]	62	Midshaft, proximal	Two-stage graft	11 (18%)	26 (mean)	0	3	3	0	0	3

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### Length of follow-up is key

# **L**

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### Summary of published outcomes

Lower urinary tract function	
Spraying	NR - 63 %
Post void dribbling	NR – 30 %
Stream deviation	NR – 26 %
LUTS	NR – 20 %
Fistulae	0 % - 25 %
Stricture	0 % - 8 %
Cosmesis	
Patient dissatisfaction (mostly size)	NR – 81 %
Surgeon satisfaction	NR – 97 %
Psychosexual	
Sexual satisfaction	NR – lower - Equal
Curvature	NR – 15 %
Erectile difficulties	NR – 73 %
Ejaculation problems	NR – 36 %

#### Compiled by P Hoebeke for 2018 EAU consultation

# Quality of data in hypospadias CONSORT criteria

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Braga et al JPUrol 2017

43

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#### Corporeal Measurements NORMAL



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Silver et al 1997

EXSTROPHY





### Exstrophy cavernosogram







#### **Corporal Anatomy**

No cross circulation

- Despite previous reconstruction
- Independent blood supply
- Allows Disassembly

**Reduced length** 

Increased diameter

Surgical distortion





### Perineal skin flap vaginoplasty

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#### Flap Vaginoplasty

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Aim to avoid using bowel if possible



#### Ileal Vaginoplasty

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#### Vaginal agenesis

Ileal vaginoplasty Long term result



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### How should we measure an outcome?





### Problems vs complications



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### Outcomes in Exstrophy

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- Orthopaedic
- Urinary
- Sexual



### Outcomes in Exstrophy

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#### Orthopaedic

Increased Incidence of back pain

Hip pain up with osteotomy

#### Urinary

90% require diversion53 % develop stonesRarely void spontaneously



Sexual

Dorsal Chordee Impact on fertility (M&F) Mod Sexual Dysfunction (M&F)

56



#### Body image in exstrophy Pennison et al 2013



#### Table 2. UBIQ question results

Question	p Value			
Avoids activities with friends	1.0			
Avoids sleeping at friend house	0.09			
Avoids wearing bathing suits	0.35			
Avoids changing clothes in front of peers	<0.0001 (BE >KS)			
Thinks private parts (penis or vagina) look different	<0.0001 (BE >KS)			
Tells friends about condition	<0.0001 (BE <ks)< td=""></ks)<>			
Worries that friends will find out about condition	0.002 (BE >KS)			
Is comfortable discussing condition with friends	0.006 (BE <ks)< td=""></ks)<>			
Thinks life in future will be different because of condition	0.91			
Thinks will be less likely to have romantic relationships because of condition	0.002 (BE >KS)			
Thinks will be less likely to have children because of condition	0.003 (BE >KS)			

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#### Sexual Outcomes Deans 2012, Rubenwolf 2016



42/52 definite sexual activity

- 2 did not
- 8 declined or info not known
- 34 (65%) Exstrophy vaginoplasty to enable sexual activity

31 % at risk of sexual dysfunction



### **L**UCL

#### **Pregnancy Outcomes**

	Singleton Pregnancies N=54	Twin Pregnancies N=3
Live Birth	31	3
Miscarriage	21	
Termination	1	
Still Birth	1	3

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All caesarian sections – 3 emergencies – assoc with 2 neonatal deaths Our neonatal death/stillbirth rate – 7/100 National data 8.5/1000

High incidence of miscarriage ? False positive tests





### thebmj Research - Education - News & Views - Campaigns

#### Editorials

#### Parental choice on normalising cosmetic genital surgery

*BMJ* 2015 ; 351 doi: http://dx.doi.org/10.1136/bmj.h5124 (Published 28 September 2015) Cite this as: *BMJ* 2015;351:h5124

Liao, Wood, Creighton, October 2015





### Mantras.....







### Surgeons cannot make 'normal'







### Surgeons cannot make 'normal'

### Primary surgery is easier than revision surgery at any age





These conditions are lifelong and affected young adults need to learn to be independent in healthcare.

Some have major surgery in childhood

We are just starting to understand the long-term outcomes

MUCH more work is needed

Multidisciplinary teams are essential for good transition.