Should Paediatric and Adolescent Gynaecology be Centralised?
Disclosure of interests:

There is no conflict of interest to declare
What does Paediatric and Adolescent Gynaecology cover?

- Depends on your country’s health system

  **UK:** Primary care → Secondary Care → Tertiary Care
  
  **USA:** Direct Access to Secondary Care → Tertiary Care
  
  **European countries:** varies
  
  **Sweden:** ?? Same as UK
Sweden is best in the world for health outcomes

HOWEVER: “Biggest challenge is the issue of coordinating care between hospitals, primary care and local authorities
What constitutes PAG Care?

- Neonates and infants
- Children
- Adolescents
- Transition to adult services and continued care
UK System

- Neonates and infants
- Paediatric endocrinology
- Paediatric urology
- Paediatric surgeons
- Geneticists

Correction of disorders of sexual development in specialist centres.

Really no input from gynaecologists
UK system

- Children

Paediatric Endocrinology, Surgery and Urology

Disorders of growth and development inc puberty in children’s specialist hospitals
UK System

- Adolescents
- Primary care
- Paediatric endocrinology
- Gynaecology - complex holistic care
Problem

• Care is now fragmented
• Multiple specialties usually in different institutions
• Consultants not specialists
• Access to specialist nurses, specialist psychology etc becomes very difficult
• Funding issues
UK System

- Transition to adult services

- Usually very poor - non-specialist doctors whether surgeons, urologists, endocrinologists or gynaecologists.

- Major lack of holistic approach to care when needed.
UK Primary Care

- Menstrual disorders
- PCOS
- Contraception
- (No cervical smears before age 25)

Only refer when unable to cope
Nearly all patients seen in general gynaecology out-patient clinics i.e. with adults. May be seen by a junior doctor.

- Menstrual disorders and Amenorrhoea
- PCOS
- Genital injuries
- Pelvic pain and endometriosis
- Congenital anomalies
- Unwanted pregnancy
UK Special Clinics

- Adolescent Gynaecology clinics
- Sexual health
- Child/Adolescent Sexual Abuse
- Female Genital Mutilation - new issue!!
- Family Planning Clinics
- Abortion Services
- Premature Menopause
USA

- Same system as UK for neonates, infants and children
- Adolescent system relies entirely on gynaecologists
- Referral rate to specialist care very low - so care very poor
- Distance is a major issue
What services should be in specialist centres?

- Specialist Gynaecologists - PAG trained
- Endocrinologist with interest in disorders of puberty, CAH, induction of puberty AND able to continue care into adulthood
- Psychologists - specially trained
- Clinical nurse specialists
- Specialist radiologists
- Geneticist if needed
- Access to reconstructive surgeons/urologists
Do all patients need to be referred?

- Absolutely not.
- Referral pathways for those conditions best cared for in the centre.
- National centre to optimize care
- Locally recognised gynaecologists to be part of a network to be involved in long term care
- National organisation to establish standards of care and monitor outcomes
- This approach delivers the best care for patients.
How does this translate in practice?
Centre for Disorders of Sexual Development and Adolescent Gynaecology

- National Centre at QCCH since 1999.
- Excludes surgical problems in children under age 12.
- Focus on holistic, multidisciplinary care.
Spectrum of problems

3 main groups

- Disorders of sexual development
  - MRKH syndrome
  - Outflow tract obstruction
  - Uterine anomalies
  - 46XY DSD
  - Turner’s syndrome
  - CAH in adolescents
  - Children with vulval abnormalities
Adolescent gynaecology

- Disorders of puberty
- Menstrual abnormalities
- Complex Contraception
• Endocrine problems
  - Primary amenorrhoea
  - Secondary amenorrhoea
  - PCO
  - Hirsutism
Multidisciplinary team

- Psychologist
- Specialist imaging expertise
- Self help group for MRKH
- Endocrinologist
- Gynaecologists
Congenital Malformations of the Genital Tract

- Classification

Numerous different attempts - all variations on the same themes

ESHRE/ESGE classification 2013
## ESHRE/ESGE classification
### Female genital tract anomalies

<table>
<thead>
<tr>
<th>Uterine anomaly</th>
<th>Sub-class</th>
<th>Cervical/vaginal anomaly</th>
<th>Co-existent class</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>U0</strong> Normal uterus</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
| **U1** Dysmorphic uterus | a. T-shaped  
   b. Infantilis  
   c. Others | | |
| **U2** Septate uterus | a. Partial  
   b. Complete | | |
| **U3** Bicornoreal uterus | a. Partial  
   b. Complete  
   c. Bicornoreal septate | | |
| **U4** Hemi-uterus | a. With rudimentary cavity (communicating or not horn)  
   b. Without rudimentary cavity (horn without cavity/no horn) | | |
| **U5** Aplastic | a. With rudimentary cavity (bi- or unilateral horn)  
   b. Without rudimentary cavity (bi- or unilateral uterine remnants/aplasia) | | |
| **U6** Unclassified malformations | | | |

### Associated anomalies of non-Müllerian origin:

### Drawing of the anomaly
• Could be useful if everyone used it and reported all findings to a central resource within ESHRE but so far not found widespread uptake.

• Any research study would benefit as all researchers would use the same classification.
MRKH

• Second commonest cause of primary amenorrhoea after Turner’s syndrome.

• Differential diagnosis

  XY DSD - Androgen insensitivity

                     absence of pubic/axillary hair
INCIDENCE

- 1 in 5000 female births
  based on Finnish study
  
  (Aittomaki et al 2001)

- No reliable data for other populations
DIAGNOSIS

- Clinical
- Imaging – Ultrasound
  MRI
- Laparoscopy is un-necessary in the majority of cases
ASSOCIATED CONGENITAL ABNORMALITIES

- Renal agenesis 30%
  - Horseshoe kidney 5-10%
  - Pelvic kidney 1%
  - Duplication of ureters

- SKELETAL ANOMALIES 12%
  - Spine 60%
  - Limb
  - Rib

- URINARY TRACT ANOMALIES 40%

- HEARING IMPAIRMENT up to 10%
Classification

- **Typical** - sole anomaly (64%)
- **Atypical** - typical + renal/skeletal/hearing or other anomalies (24%)

**MURCS** - Müllerian aplasia, renal aplasia and cervicothoracic somite dysplasia (12%)

Oppelt et al 2006
PSYCHOLOGICAL FACTORS

- SHOCK
- DEPRESSION
- DOUBTS OF GENDER
- INFERTILITY
- SEXUALITY
- WORTHLESSNESS
- CULTURAL DIFFICULTIES
PARENTAL PROBLEMS

- DEPRESSION
- FEAR
- IGNORANCE
- ACCEPTANCE OF INFERTILITY
- PATERNAL SUPPORT
IMPORTANCE OF PSYCHOLOGICAL PREPARATION

- Dealing with adolescent stress and difficulties
- Dealing with sexuality
- Dealing with support mechanisms
- Success of interventions is directly related to psychological success
Surgical Management of MRKH Syndrome

- Vulvoplasty  
  William’s operation

- Bowel Vaginoplasty  
  Ileum  
  Sigmoid colon  
  Caecum

- Vecchetti’s Operation  
  Laparotomy  
  Laparoscopic
## Success of Surgical Techniques

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Success Rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vulvoplasty</td>
<td>95%</td>
</tr>
<tr>
<td>Vaginoplasty Amnion</td>
<td>84%</td>
</tr>
<tr>
<td>McIndoe</td>
<td>92%</td>
</tr>
<tr>
<td>Davidov</td>
<td>88%</td>
</tr>
<tr>
<td>Sigmoid</td>
<td>88%</td>
</tr>
<tr>
<td>Vecchetti</td>
<td>95%</td>
</tr>
<tr>
<td>Buccal Mucosa</td>
<td>90%</td>
</tr>
</tbody>
</table>
NON-SURGICAL MANAGEMENT

VAGINAL DILATORS

- Repeated use of graduated vaginal dilators
- Careful instruction
- 3 Times daily for 20 minutes for 2-3 months
Various Vaginal Dilators
## Results of Dilator Therapy

<table>
<thead>
<tr>
<th>Author</th>
<th>Patient Nos</th>
<th>Success</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rock et al (1983)</td>
<td>21</td>
<td>18 (86%)</td>
</tr>
<tr>
<td>Broadbent et al (1984)</td>
<td>20</td>
<td>19 (95%)</td>
</tr>
<tr>
<td>Roberts et al (2001)</td>
<td>51</td>
<td>46 (91%)</td>
</tr>
<tr>
<td>Gargollo et al (2009)</td>
<td>57</td>
<td>50 (88%)</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>149</strong></td>
<td><strong>133 (89%)</strong></td>
</tr>
</tbody>
</table>
Results of Dilator therapy

- Edmonds et al 2012

- Experience of 245 consecutive patients with MRKH over 12 years

  232 (95%) anatomical/functional success

- 13 did not complete therapy:
  - Psychiatric or cultural issues only

- Therefore 100% success if therapy completed.
Updated results to 2015

- Total number of patients completing dilator program: 330
- Success rate: 100%
MRKH SURROGACY

- Widely accessed
- No international data base
- Few reported series
- Success similar to IVF
- No reported female offspring with MRKH
Uterine transplantation

- Controversial
- Ethical issues re non-life saving transplantation
- Costs within a nationally funded health system and competing resources
- Long term health issues for recipient, donor and offspring remain unknown
IMPERFORATE HYMEN

Bulging and “blue”
Management

- Incision and drainage
- No sequelae
TRANSVERSE VAGINAL SEPTUM

- CYCLICAL ABDOMINAL PAIN
- PRIMARY AMENORRHOEA
- SECONDARY SEXUAL CHARACTERISTICS PRESENT
MRI
Levels of Obstruction

- a – high
- b – middle
- c – low
Surgery

- Excision of septum and vaginal advancement
- Post operative dilators/mould
OUTCOME

• Sexual function

  10% dyspareunia rate for low septae

  40% for high septae

• Endometriosis

• Reproductive performance

  100% for low problems

  20% for high obstructions
LONGITUDINAL VAGINAL SEPTUM

- DOUBLE VAGINA
- OBSTRUCTED HEMI-VAGINA
DOUBLE VAGINA

- DIFFICULTY USING TAMpons
- DYSPAREUNIA
- ANTE-NATAL DIAGNOSIS
- INTRA-PARTUM DIAGNOSIS
Management

• Excision surgery
  Ligation
  Diathermy
  Laser
  Harmonic scalpel

• Complications
  Haemorrhage, infection
  Dyspareunia
Need for a Swedish Society of Paediatric and Adolescent Gyneacology

- Education
- Training
- Research
- Communication - Professional, patients, parents
- Information
- Clinical standards
- Professional Opinion
Current Issues

- Female Genital Mutilation (FGM)
- Labial reduction
- Sexual abuse
- Childhood and adolescent obesity
- Athletic Triad
- Onco-fertility