PAEDIATRICS

# A NATIONAL MODEL OF CARE FOR PAEDIATRIC HEALTHCARE SERVICES IN IRELAND

## CHAPTER 28: PAEDIATRIC GYNAECOLOGY

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# 28.0 INTRODUCTION

The management of young children and adolescents with gynaecological problems requires sensitivity in relation to their specific needs. This extends from immediate service provision in acute cases to the more long term management of complex issues relating to sexual function, psychology, endocrinology and fertility. The specialty manages children aged 0-16 years inclusive, and appropriate transition arrangements are essential.

*The main conditions seen are:*

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| **General Paediatric Gynaecology Conditions** | Premenarcheal bleeding<br>Recurrent vulvovaginitis<br>Labial adhesions<br>Vulval skin conditions, e.g. lichen sclerosus<br>Genital tract abnormalities<br>Menstrual dysfunction |
| **Disorders of Puberty and Adolescence** | Delayed puberty & variants<br>Hypoestrogenic states leading to absent menstruation<br>*e.g. eating disorders, intensive exercise, hyperprolactinaemia*<br>Premature ovarian failure<br>Precocious puberty & variants<br>Polycystic ovary syndrome<br>Adolescent menstrual disorders and pelvic pain<br>Gynaecological aspects of late effects of cancer |
| **Developmental Anomalies** | Mullerian Anomalies:<br>• Agenesis e.g. Absence of the uterus and upper vagina, as in the Mayer-Rokitansky-Kuster-Hauser Syndrome (MRKH)<br>*or*<br>• Partial or total duplication of the uterus, cervix and vagina, in which cases presentation may be in adolescence with menstrual obstruction or in adult life with miscarriage and subfertility<br>• Obstruction, as in transverse vaginal septae, cervical agenesis and some obstructive duplication anomalies<br><br>Disorders of Sex Development (DSD):<br>• 46 XY DSD including disorders of gonadal development such as Swyer Syndrome and disorders of androgen synthesis or action such as Androgen Insensitivity Syndrome (AIS), 5-alpha reductase deficiency and 17 hydroxysteroid dehydrogenase deficiency<br>• 46XX DSD including disorders of gonadal development such as gonadal dysgenesis as well as androgen excess, e.g. Congenital Adrenal Hyperplasia<br>• Sex chromosome DSD including Turner syndrome and variants and 46X/46XY mixed gonadal dysgenesis |

Complex urological anomalies such as cloacal and anorectal malformations have a high incidence of associated congenital abnormalities. There are also a small number of young women with complex acquired gynaecological abnormalities who require a similar multidisciplinary approach.

# 28.1 CURRENT SERVICE PROVISION

The service is run by a single gynaecologist and is largely an outpatient service. There is one clinic per week (46 weeks per year), and two additional clinics per month, with 10-15 attendances per clinic. There were 327 children and adolescents seen in 2013, with 207 new referrals. There are strong links with urology/general surgery/ adult gynaecology, and links to St. Clare's child sexual assault (CSA) assessment unit, Temple Street Children's University Hospital with 3-5 examinations per month.

*The most common diagnoses seen are:*

- Vulval discharge
- Abnormal appearances of the vulva
- Hymen issues
- Menorrhagia or amenorrhoea

*Current service deficits have been identified as follows:*

- Single-handed service
  The service cannot develop with a single-handed gynaecologist, and an expansion to 2 x 0.5WTE in child and adolescent gynaecology is required.
- No on-call service
  On-call issues do arise, and an on-call service is required.
- No gynaecology colleague in Our Lady's Children's Hospital, Crumlin
  This means that all cases need to be referred to Temple Street.
- Need stronger links with urology (especially for cloacal abnormalities)
  Current deficits in paediatric urology need to be addressed.

# 28.2 PROPOSED MODEL OF CARE

*There are two types of presentation:*

1. Presentation at birth or in childhood with a DSD (commonest presentation is with ambiguous genitalia) or complex urological anomaly. These patients will require transition from paediatrics for specialised adolescent and adult services.
2. New diagnosis in adolescence, e.g. primary amenorrhoea with or without pubertal development, virilisation at puberty, or obstructed menstruation. These patients will present to their general practitioner (GP) who will usually refer them to the specialist adolescent gynaecology service.

### Service Description / Care Pathway

Services will provide the defined activities outlined below as part of a multidisciplinary team approach associated with interdependent services.

*The patient journey will include:*

| | | | | |
|---|---|---|---|---|
| Referral from GP or secondary care | Investigations and diagnosis | Inpatient or outpatient treatment | Outpatient follow up | Discharge where appropriate |

*The multi-disciplinary team will include:*

- Co-located:
  - Paediatric gynaecologist
  - Psychologist
  - Specialist nurse
- Interdependent:
  - Endocrinologist, urologist, biochemist, geneticist, plastic surgeon, radiologist as required

*Management of congenital anomalies of the genital tract includes two main diagnostic groups:*

1. Disorders of sex development
   Older terminology now abandoned by clinicians (but still present in ICD classification) includes intersex, indeterminant sex and hermaphrodite/pseudohermaphrodite.

2. Complex mullerian anomalies
   This includes Rokitansky syndrome (mullerian agenesis) and mullerian obstruction/duplication. Some complex mullerian anomalies are associated with complex urological anomalies such as bladder extrophy and cloacal anomaly.

*Outpatient Services*

All patients will require assessment by a multidisciplinary team. The composition of the team may vary slightly depending on diagnosis. Patients with complex mullerian anomalies will need to see a gynaecologist, clinical nurse specialist and psychologist. Gynaecology input should include access to advanced minimal access surgery. Those with a DSD will also need specialist endocrinology and urology input. The service must be supported by specialised imaging, biochemisty and genetics. Family members may need genetic referral.

At least two appointments will be required. The second appointment will include review of results and disclosure of information including atypical karyotype. Further psychology input is usually required for this in the majority of cases.

*Investigations*

Complex imaging should include magnetic resonance imaging (MRI). Genetics investigations will include karyotype but also specialised genetic testing in the case of some DSDs, e.g. androgen insensitivity syndrome.

*Treatment Strategy*

Treatment is diagnosis dependent and may be non-surgical (dilation or hormone replacement) or surgical:

- Non-surgical dilation for neovaginal creation is the first line treatment of choice for vaginal agenesis and is successful in 85% of cases. It is a nurse led treatment but requires contact with the patient (face-to-face or telephone) every two to three weeks for approximately three to six months.
- Surgical treatment will vary due to the diagnosis and previous surgical procedures. Procedures should be performed laparoscopically. Patients with previous vaginal or abdominal reconstructive surgery may require vaginoplasty with skin flaps or intestinal vaginal replacement. These procedures should be performed jointly with specialised urology.

Written age-appropriate information should be available for the commonest conditions. Information leaflets should be family-orientated.

### Specialist Centre

All clinicians involved in the care of children with complex PAG conditions should have access to a specialist centre. This centre should have the expertise to manage the full range of DSDs within a complete multi-disciplinary service. A hub and spoke arrangement is required between regional and local hospitals and the specialist referral centre.

The centre should be able to manage most complex developmental anomalies, such as anomalies of the mullerian tract and fusion abnormalities of the vagina. Initial examination may need to include specialised imaging such as 3D ultrasound and MRI. Available treatments should include dilator training for congenital absence of the vagina and reconstructive surgery for the more complex mullerian tract anomalies, congenital adrenal hyperplasia and other rare disorders of sex development. Surgery may be complex and expert urology input should be available.

Developmental anomalies are generally more complex and require appropriate recognition and diagnosis. Many need rapid referral and access to the specialist centre. A paediatric and adolescent gynaecologist should, however, be able to manage simple vaginal fusion anomalies such as imperforate hymen and vertical septa with additional training being required for the treatment of transverse vaginal septa and more complex anomalies.

Psychological input is an integral part of multidisciplinary care both during investigation and treatment and should be easily accessible. This should focus on the direct social and emotional consequences of the diagnosis and treatments. There should be at least one specialist nurse and an identified clinical psychologist.

Complex patients will need to be seen in combined clinics with consultants in paediatric and adult endocrinology and paediatric surgery, depending upon clinical throughput. It is expected that the full range of multi-disciplinary support be available including specialist diagnostics (radiology, clinical biochemistry, haematology, and genetics), plastic surgery, psychology and support groups with access to mental health and social care as needs arise. The specialist centre is expected to maintain links with patient peer support groups and co-ordinate and contribute to research in PAG.

### Transition to Adult Services

Transfer of adolescents with a known diagnosis into the adult service can be haphazard. There should be close links between the adult and the paediatric services, preferably with transition clinics held jointly between the paediatric and adult teams. Adolescents with DSD will require information and counselling on sexual health, contraception and fertility. The parents of younger children often request information on fertility potential.

## 28.3 REQUIREMENTS FOR SUCCESSFUL IMPLEMENTATION OF MODEL OF CARE

### Infrastructure

- Appropriate examination room
- No inpatient bed requirements
- Accommodation for twice weekly outpatient clinics
- Colposcope

### Staffing

There should be an identified consultant gynaecologist as lead clinician, who should ideally have developed a special interest in paediatric and adolescent gynaecology (PAG), and be a member of The British Society for Paediatric & Adolescent Gynaecology and attend regular meetings and updates. Practitioners need to able to recognise cases of child sexual abuse and sexual assault as part of the differential diagnosis and be aware of the appropriate local referral pathways.

| Current | Recommended |
|---|---|
| 0.2WTE paediatric and adolescent gynaecologist | 1.0WTE paediatric and adolescent gynaecologist<br><br>2WTE clinical nurse specialist (CNS) in paediatric and adolescent gynaecology<br><br>Access to psychology services in specialist centre |

### Education and Training

Development of the CSA assessment service should be separate to the adolescent gynaecology service. Evolution of the speciality will require collaboration with UK/Australian/US colleagues and training fellowships abroad.

### Interdependencies with Other Services

- Teenage pregnancy clinics with separate antenatal education and a specialist nurse (current service has 100-200 13-17 year olds per annum)
- Strengthen links with sexually transmitted infections services in St James's Hospital
- There should be paediatric endocrinology input for cases relating to precocious or delayed puberty and Turner's syndrome

## 28.4 PROGRAMME METRICS AND EVALUATION

*The British Society for Paediatric and Adolescent Gynaecology recommends the following core standards:*

- There should be a designated lead clinician for PAG.
- Care (whether for general PAG or complex cases) needs to be in an appropriate setting with facilities for outpatient and inpatient management of children, adolescents and their families.
- Children with gynaecological problems should not be seen in the setting of an adult gynaecology clinic. Designated regular PAG clinics supported by a paediatric and/or specialist nurse should be established in units where children with gynaecological problems are expected to be assessed and treated.
- The lead clinician should be the first point of contact where possible in cases of acute gynaecological conditions involving children. Admissions should be to a paediatric ward.
- All healthcare professionals involved in the management of these cases require child protection training appropriate to their roles.
- The specialist centre needs to provide a multidisciplinary approach to treatment. This includes joint clinics and theatre sessions with paediatric surgeons, urologists (often paediatric and adult), plastic surgeons, paediatric and adult endocrinologists and additional input from geneticists, radiologists, specialist nurses, psychologists and psychiatrists as necessary.

## 28.5 KEY RECOMMENDATIONS

- There should be a designated lead clinician for paediatric and adolescent gynaecology. An expansion to 1WTE consultant is required.
- In a specialist centre, there should be at least one specialist nurse and an identified clinical psychologist.
- Transition clinics should take place between paediatric and adult services.

## 28.6 ABBREVIATIONS

| | |
|---|---|
| CNS | Clinical Nurse Specialist |
| CSA | Child Sexual Assault |
| DSD | Disorders of Sex Development |
| GP | General Practitioner |
| MRI | Magnetic Resonance Imaging |
| PAG | Paediatric and Adolescent Gynaecology |
| WTE | Whole Time Equivalent |

## 28.7 REFERENCES

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