The British Society for Paediatric & Adolescent Gynaecology.

Clinical Standards for Service Planning in PAG

Introduction

The management of young children and adolescents with gynaecological problems (aged 0-18 years inclusive, with appropriate transition arrangements) 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 British Society for Paediatric & Adolescent Gynaecology (the Society) is a multidisciplinary Society which was created in 2000 and which has sought to develop a network of Clinical Centres where children and adolescents with gynaecological problems can be seen, assessed and managed to a high standard.

The Department of Health has recently acknowledged the need for a network of specific centres to deal with complex cases relating to the management of Disorders of Sex Development (DSD) as well as other complex and rare cases. The age of presentation of DSD will vary depending on the type of problem and the timing of its manifestation and can occur within the spectrum ranging from the neonatal period to late adolescence and beyond. The prevalence of other congenital anomalies present varies depending upon the population studied and the age at which the problem is likely to be detected.

Core Principles

The Society believes it to be imperative that:-

(a) Acute Hospital Trusts providing Gynaecological services provide a designated Consultant-led service for children with Gynaecological problems and seek to provide a dedicated service to these children.

(b) Disorders of Sex Development (DSD) are managed in Specialist Centres by a multi-disciplinary team. This includes paediatric surgeons, urologists, plastic surgeons, endocrinologists, specialist nurses, psychologists and also the gynaecologist, whose role is to help co-ordinate care, and in particular the transition from childhood through to adolescence and womanhood, helping with issues relating to sexual function, psychology, endocrinology and fertility.

The main conditions that are covered in the scope of this paper are:-

(i) General PAG Conditions

To include:

- *Premenarcheal bleeding*
- *Recurrent vulvovaginitis*
- *Labial adhesions*
- *Vulval skin conditions e.g. lichen sclerosus*
- *Genital tract abnormalities*
- *Menstrual dysfunction*

(ii) Disorders of Puberty and Adolescence

To include:

- *Delayed puberty & variants.*
- *Hypoestrogenic states leading to absent menstruation e.g. eating disorders, intensive exercis*e, *hyperprolactinaemia.*
- *Premature ovarian failure*
- *Precocious puberty & variants*
- *Polycystic ovary syndrome*
- *Adolescent menstrual disorders and pelvic pain*
- *Gynaecological aspects of late effects of cancer*

(iii) Developmental Anomalies

To include:

- *Mullerian Anomalies*
    - *Agenesis e.g. Absence of the uterus and upper vagina, as in the Mayer-Rokitansky-Kuster-Hauser Syndrome (MRKH)*
      or
      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.
    - *Obstruction, as in transverse vaginal septae, cervical agenesis and some obstructive duplication anomalies*
- *Disorders of Sex Development (DSD)*
    - *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*
    - *46XX DSD including disorders of gonadal development such as gonadal dysgenesis as well as androgen excess e.g. Congenital Adrenal Hyperplasia.*
    - *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. This includes vaginal loss secondary to trauma, or else due to radical surgery/irradiation for childhood genital tract malignancies.

Patients may be referred directly by their primary care physician to the gynaecology service or may be referred by colleagues in other disciplines.

Referrals may result from pre-existing conditions that require review or new problems arising in childhood, puberty and adolescence.

There is little information on the true prevalence of many of the conditions referred to within this document. The outcomes of treatment of complex cases can be difficult to quantify in the long term.

Core Standards

Despite the recognised difficulties in defining auditable standards this is an area which The British Society for Paediatric and Adolescent Gynaecology is attempting to develop.

The following are the Society's defined core standards:-

- ✓ There should be service provision for children and adolescents with gynaecological problems in all acute hospitals providing gynaecological services. Each unit should have a designated Lead Clinician for PAG. The provision of a Lead Clinician within each unit improves patient care[1]

- ✓ 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[2,3].

- ✓ 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 Level 1-2 Child Protection Training depending on their roles, and access to study leave to attend meetings in order to obtain regular updates. The Lead Clinician should have received Level 3 training.

- ✓ Clinical networks should be developed flexibly on a geographical basis to allow for support to Lead Clinicians and the transfer of care of complex cases from secondary/general PAG care to Specialist Centres. Appropriate onward referrals can then be made for those complex cases after initial work-up.

- ✓ There should be a limited number of designated Specialist Centres for the management of the very rare and more complex conditions.

- ✓ These Specialist Centres need 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.

- ✓ Training facilities must be available at both General and Specialist Centre levels in order to provide basic training and support trainees who may be undertaking the Advanced Training Skills Module in PAG or Subspecialty Training in Reproductive Medicine.

- ✓ Each Centre is encouraged to maintain its own database of cases managed locally and provide data for local Congenital Abnormality Registers.

- ✓ The Society will maintain a database of regional networks and details of local PAG service facilities to be made available on the BritSPAG website accessible to all.

(i) General PAG Conditions.

There should be a designated local service available for the management of general PAG problems[4].

The provision of dedicated universal general PAG clinics in Acute Hospital Trusts offering Gynaecological services is a key desirable outcome for the Society.

This will allow General Practitioners to obtain specialised advice in areas where there is often little experience and will encourage education and training of both established clinicians and trainees.

There should be an identified Consultant Gynaecologist as Lead Clinician, who should ideally have developed a special interest in PAG and may have obtained the Special Skills Module, ATSM or hold Subspecialty training in Reproductive Medicine.

The Lead Clinician should ideally be a member of The British Society for Paediatric & Adolescent Gynaecology and attend regular meetings and updates.

The Lead Clinician should be supported by a trained paediatric and/or specialist nurse (at least Band 5/6) who is likely to be based in the Outpatients Department but may be Ward based, depending upon the size of the Department.

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.

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

(ii) Disorders of Puberty and Adolescence

These conditions are relatively common and should be managed by all Acute Hospital Trusts providing gynaecological and paediatric services. There should be paediatric endocrinology input for cases relating to precocious or delayed puberty and Turner's syndrome, with induction of puberty managed by a Consultant with a special interest, likely to be based in a Specialist Centre. Some Specialist Centres hold Turner's syndrome clinics in order to review patients on an annual basis. There is an overall need for adequate provisional of transitional care from paediatric to adult endocrine services.

Lead clinicians should develop a regular dedicated PAG clinic and children should not be seen in adult general gynaecology clinics.

A hub and spoke arrangement is required with the local specialist referral centre. This will allow for input from sub-specialists in reproductive medicine for the more complex patients.

Appropriate advice and protocols can be provided from the Specialist Centres as necessary.

(iii) Developmental Anomalies

These conditions are generally more complex and require appropriate recognition and diagnosis. Many need rapid referral and access to the nearest and most appropriate specialist centre.

The Lead Clinician 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.

Specialist Centres

All clinicians involved in the care of children with complex PAG conditions should have access to Specialist Centres.

Specialist Centres should have the expertise to manage the full range of Disorders of Sex Development within a complete multi-disciplinary service.

Clinical networks may need to link between more than one specialist centre. In addition there may be occasional extremely complex cases that require management in only one or two specific centres in the UK offering the required level of care over and above recognised specialist centres.

Specialist centres 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. Some procedures such as gonadectomy and removal of an obstructed uterine remnant can now be carried out laparoscopically and minimal access surgery should be considered as the preferred approach. Intestinal vaginal substitution may be needed and may require access to urology, plastic surgery and colorectal surgery. 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 e.g. managing self-disclosure, overcoming social avoidance and sexual activity, and generating realistic treatment expectations. Where generic mental health problems are identified, the usual community resources should be accessed and the psychologist will assume the liaison role for the team.

There should be at least one specialist nurse (at least Band 5/6) and an identified healthcare 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[5]. Some decisions may require the involvement of a medical ethicist.

Other than for the management of common and complex PAG conditions, Specialist Centres are expected to maintain links with patient peer support groups and co-ordinate and contribute to research in PAG

Transfer of adolescents with a known diagnosis into the adult service can be haphazard and 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. There should be access to a Reproductive Medicine Subspecialist, who may continue care into adulthood.

Acknowledgements:

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References:

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Chair BritSPAG, January 2011