

SCHEDULE 2 – THE SERVICES

A. Service Specifications

Service Specification No.	E10c (merged with former E10s(HSS)/b)
Service	Congenital Gynaecological Anomalies
Commissioner Lead	
Provider Lead	
Period	
Date of Review	

<p>1. Population Needs</p>
<p>National/local context and evidence base</p> <p>Complex Congenital Gynaecological Anomalies refers to the gynaecological management of rare congenital anomalies of the genital tract. The majority of cases present in one of two ways – either at birth or at adolescence. Some cases that present in childhood or adolescence will need long-term specialised gynaecological management into adulthood.</p> <p>Patients who present at birth with these conditions are managed by a specialist paediatric team. They are transferred to an adolescent/adult team when judged suitable. The timing of this depends on the medical requirements and maturity of the individual patient as well as the local service configuration. Those patients who are newly diagnosed in adolescence are referred directly to an adolescent gynaecologist and will not usually see a paediatrician.</p> <p>Evaluation and long-term management must be performed at a centre with an experienced multidisciplinary team^{1,2}. Multi-professional and multi-disciplinary input is required either because the patient has associated co-morbidities which render the care of her gynaecological disorder especially complex e.g. endocrine problems such as Congenital Adrenal Hyperplasia or a complex urological condition (such as cloacal anomaly) or because she may require complex treatment including reconstructive surgery to optimise future gynaecological, psychological and reproductive outcomes. This is best provided by a specialist centre co-located with other specialised services such as paediatric gynaecology, urology, endocrinology, psychology, radiology and genetics.</p> <p>Units that provide this service will also need to define links to other services outlined in section 3.2. The Provider will deliver high quality services within an agreed network of Providers whose population may also use this service.</p> <p>Close links with services providing care for children with these complex conditions are essential. Some of these conditions will require lifelong medical care. Transition from paediatric to</p>

adolescent to adult services must be seamless and poor transition has been implicated in non-compliance with follow-up. Gynaecologists working within these services will manage both adolescent and adult patients ensuring smooth transition to adult services.

Complex Congenital Gynaecological Anomalies includes the following two diagnostic groups;

- Disorders of Sex Development (DSD). Older terminology now abandoned by clinicians (but still present in ICD classification) includes Intersex, Indeterminant sex and hermaphrodite/pseudohermaphrodite.
- Complex Mullerian Anomalies. This includes Mayer-Rokitansky--Küster-Hauser (MRKH) syndrome (mullerian agenesis, MRKH) and mullerian obstruction/duplication. Some complex mullerian anomalies are associated with complex urological anomalies such as bladder extrophy and cloacal anomaly.

Accurate incidence figures are not available but these conditions are estimated to affect 1 in 2-3000 females. A review of the incidence of the most commonly seen conditions gives an indication of the expected numbers (Appendix). This is in the order of 25 000 cases in the current population of UK women.

2. Outcomes

2.1 NHS Outcomes Framework Domains & Indicators

Domain 1	Preventing people from dying prematurely	√
Domain 2	Enhancing quality of life for people with long-term conditions	√
Domain 3	Helping people to recover from episodes of ill-health or following injury	√
Domain 4	Ensuring people have a positive experience of care	√
Domain 5	Treating and caring for people in safe environment and protecting them from avoidable harm	√

Outcome measures:

These will be determined annually by national peer review via the British Society for Paediatric and Adolescent Gynaecology (BritSPAG) and national standards for paediatric and adult care. Clinical outcomes will be monitored and include:

- Paediatric and adolescent patients to be seen within dedicated paediatric clinics and in-patient wards
- Caseload of referrals for complex reconstructive surgery
- Proportion of reconstructive procedures performed laparoscopically
- Psychology support offered to all patients

- Outcome of vaginal dilation treatment for vaginal agenesis

3. Scope

3.1 Aims and objectives of service

The aim of the service is to provide patient centred specialist care for adolescents and adult women with a congenital abnormality of their genital tract and promote family centred specialist care and to optimise future reproductive and sexual function as well as psychological health and quality of life.

The primary aims are:

- To provide a safe and effective care pathway for adolescent and adult women with congenital anomalies of the genital tract.
- To provide social, economic and psychological benefits for adolescents and adult women and their family requiring the service
- To provide continuity of care through the whole care pathway encompassing other specialised services included within the pathway including paediatric and adult services
- To provide a service acceptable to girls and women in which they feel comfortable and empowered to contribute to and make decisions about their own health.

3.2 Service description/care pathway

Services will provide the defined activities outlined below as part of a multidisciplinary team approach associated with interdependent services. Patients will follow a care pathway as described below and in the appendix.

The patient journey will include:

- Referral from GP or secondary care or transfer at adolescence from specialised paediatric services.
- Initial outpatient assessment
- Investigations and diagnosis
- In patient or outpatient treatment
- Outpatient follow up
- Discharge where appropriate
- Long-term management where necessary

Adolescent patients will be seen in a dedicated paediatric and adolescent gynaecology clinic located within paediatric services in age appropriate surroundings. If in-patient admission for surgery is required this will be to a paediatric ward.

All clinicians involved in the care of children should have training in safeguarding to level 3.

The multi-disciplinary team will include:

Co-located

- Gynaecologist (including advanced minimal access surgery)
- Psychologist
- Specialist nurse
- Endocrinologist
- Urologist
- Radiologist
- Paediatric services should usually be co-located but in some areas may be in close proximity depending on hospital configuration

Interdependent

- Colorectal
- Biochemist
- Geneticist
- Plastic surgery, as required
- Paediatric endocrinology
- Paediatric urology/surgery
- Paediatric psychology

Outpatient Appointment

All adolescent and adult patients will require assessment by a multidisciplinary team. The composition of the team may vary slightly. 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, biochemistry and genetics. Family members may need genetic referral and the team should co-ordinate this.

At least two appointments will be required to determine the diagnosis. 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. If surgery is required, it will be planned at this visit. Age appropriate informed consent is mandatory and psychological support for the patient and family may be required to facilitate this.

Investigations

As listed. Complex imaging will include MRI. Genetics will include karyotype but also specialised genetic testing in the case of some Disorders of Sex Development (DSD) (e.g. androgen insensitivity syndrome).

Treatment Strategy

- Treatment is diagnosis dependent and may be non-surgical (dilation or hormone replacement) or surgical. Psychological support must be available for all patients regardless of mode of treatment.
- Non-surgical dilation for neovaginal creation is the first line treatment of choice for vaginal agenesis and is successful in over 80% of cases. The service must be flexible and offer patient choice as to timing of dilation. Timing of treatment is an individual choice and adolescent girls may choose to commence treatment immediately or defer until adulthood. It is a nurse led treatment but requires discussion with the patient (face-to-face or telephone) every 2-3 weeks for approximately 3-6 months. Psychological support is valuable both in preparation for dilation and support for those struggling with compliance. In-patient admission to hospital for dilation has not been demonstrated to give better results and most units will offer outpatient treatment only.
- Girls and women who fail dilation or are unsuitable for dilation due to scarring or genital configuration, will require surgical vaginal reconstruction. The type of operation chosen will depend on the patient's past surgical history and the preference of the surgeon. There are no good comparative outcome studies of the various techniques available. In the absence of previous genital surgery, the laparoscopic Vecchietti procedure has been demonstrated to have a high success rate and low morbidity. However in units where minimal access surgery is not available or in cases where the patient has genital scarring or unsuitable anatomy, a variety of other surgical reconstructive procedures are used. These include vaginal reconstruction with skin flaps, skin grafts and intestine as well as the laparoscopic Davidov procedure. These procedures are usually performed as joint reconstructive cases with input from other surgical specialties such as urology, colorectal or plastics. Centres managing this group of complex patients must either be able to offer these alternative procedures or have close links with centres that can do so.
- Surgical treatment for other complex anomalies will vary due to the diagnosis and previous surgical procedures. Minimal access surgery carries significant advantages in this young group of patients in terms of physical appearance and speed of recovery, meaning less time missed from school/college.. Procedures such as gonadectomy, excision of obstructed uterine horn, uterovaginal anastomosis and excision of some high transverse septums 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 or colorectal input. Plastic surgery input may be required.
- Clitoromegaly is a feature of some DSD diagnoses and can cause distress. Initial assessment and management is psychological. If surgery is required it should be performed by surgeons proficient in this procedure. Informed consent should include discussion of potential detrimental impact on sexual sensitivity and sexual function.
- Some adolescent girls will require induction of puberty and this should be led by an endocrinologist. This entails regular follow-up initially frequently and then on an annual basis. Regular bone density monitoring will be required over the age of 18. Investigation of delayed puberty should be done jointly with an endocrinologist. Management of patients with complex endocrinology such as Congenital Adrenal Hyperplasia should be led by an endocrinologist within the setting of the multidisciplinary team.
- Fertility advice and onward referral to the patient's own General Practitioner may be required in some conditions for example IVF.
- Where endocrine input is required, the transition years (from 13 to 25) should involve a paediatric and adult endocrinologist.
- Centres should develop and make available educational materials to support

understanding of treatment option and promote compliance.

Follow-up

- Follow-up for non-surgical dilation is described above.
- In addition some adolescents and adults women will be required to perform vaginal dilation after surgical vaginoplasty to prevent restenosis. This will require follow-up visits for supervision with the nurse specialist.
- Other surgical procedures such as excision of an obstructed uterine horn will require a single post-operative visit only.
- Patients with a DSD will require long-term surveillance for chronic disease. This will mean life-long specialist follow-up. Depending on the disease, either an endocrinologist or gynaecologist will be the lead clinician.

Further investigations/treatment

As in pathway. Appendix 2

Second treatment episode

- If dilation fails, surgical vaginal reconstruction is required. There are no comparative studies of the many available operative procedures. The laparoscopic Vecchietti procedure is offered in several UK units. It is widely used in Europe and research has confirmed low morbidity and high success rates. Other operations include vaginoplasty with skin graft (McIndoe-Reed procedure), rotational skin flaps and intestinal replacement. These procedures have a higher associated morbidity but are used when the Vecchietti procedure is unavailable or contraindicated.
- Vaginal stenosis is a recognised medium and long term risk of vaginoplasty and may require further treatment with dilation or surgery.

Pregnancy

1. Pregnant women with some of the pre-existing conditions as discussed in this specification e.g. Congenital Adrenal Hyperplasia, Complex Urological Anomalies require assessment and/or management from specialised maternity care delivered within a dedicated multidisciplinary service staffed by a maternal medicine specialist, a physician and supporting multidisciplinary team with experience of managing the condition in pregnancy
2. In view of this, nationally commissioned condition specific services must have outreach arrangements with specialised tertiary maternity units with access to appropriate tertiary medical, surgical, fetal medicine, clinical genetics and level 3 neonatal intensive care services. These specialised maternity services must have a critical mass of activity to maintain expertise, ensure best practice, training opportunities and for the organisational infrastructure, staffing, facilities and equipment to be clinically and economically efficient. They should have robust risk management and performance monitoring processes.
3. All such women must receive personalised pre-pregnancy and maternity planning from specialised maternity services to allow optimal disease management in the context of pregnancy. This will reduce avoidable morbidity, mortality and unnecessary intervention for

mother and baby.

4. Women with conditions discussed in this specification must be referred immediately once they are pregnant to plan their care. This must include access to termination of pregnancy and specialist advice re contraception. The individualised care plan must cover the antenatal, intra-partum and post natal periods. It must include clear instructions for shared care with secondary services, when appropriate including escalation and transfer protocols and clear guidelines for planned and emergency delivery.

Figure 1. The Pathway

See Appendix 2

Data Management , Audit and Governance

- Centres will be responsible for their clinical governance and collaborate in a quality improvement programme.
- Centres should collaborate with clinical networks established by the British Society for Paediatric and Adolescent Gynaecology.
- Clinical outcomes should be monitored as part of a peer review process under the guidance of the British Society for Paediatric and Adolescent Gynaecology.
- Within centres there should be a clear clinical governance structure for this service which involves all professionals and specialists contributing to the management of this complex group. Specific attention should be paid to the transition of adolescent patients to ensure safe and comprehensive transfer of care.

Service model

- The Service will provide a tertiary service to support adolescent girls and women requiring specialist support within a network of care and pathways.
- Close links with paediatric services providing care for children with complex disorders are essential to ensure smooth transition of girls from paediatric to adolescent to adult services. Ideally these should be co-located.
- Older adolescents may prefer to be on an adult ward and should be offered the choice.
- There will be an agreed planned and mapped pathway of care for girls and women with a complex gynaecological problem
- Gynaecologists will manage both adolescent and adult women with these conditions. As the surgical procedures are identical regardless of age, this will ensure adequate surgical activity to maintain skills.
- The Service will be part of a multidisciplinary team working together, networking and linking with other healthcare services across both community and hospital settings.
- The Service will be expected to use evidence based approaches and to demonstrate efficiencies whenever possible.
- Eligible adolescent girls and women will be referred using a defined referral system that can be audited for waiting times
- A discharge plan will be prepared offering support and facilities required providing care at home.

- Appropriate referrals to specialist colleagues will be documented and GP informed of any transfer of care.
- It is the responsibility of the Provider to recruit/provide suitable and appropriately competent and qualified personnel in the provision of this service.
- Expert psychological input targeting emotional and social adjustment is crucial. Patients are likely to require interventions to cope with an atypical karyotype and the potential emotional challenges of infertility, complications related to sexual function, orientation and identity, communication barriers within the family, friends, partner and problematic transitions.
- Surgery will be performed by gynaecologists experienced in these procedures and where appropriate with input from other specialist surgical services such as urology, plastics and colorectal.

Referral processes and sources

The service will accept referrals from primary or secondary care clinicians in adult or paediatric services as well as referrals from specialist paediatric services. Patients with congenital anomalies will normally be under the care of a gynaecologist with appropriate specialist qualifications.

The service will accept referrals from other providers particularly where the referring service is not accredited to undertake the clinical role the patient requires.

There are two types of presentation;

- 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.
- New diagnosis in adolescence. E.g. primary amenorrhoea with or without pubertal development, virilisation at puberty, obstructed menstruation. These patients will present to their GP who will usually refer them to local gynaecology services.

The Provider will be expected to use evidence based approaches and to demonstrate efficiencies whenever possible.

- Eligible patients will be referred using a defined referral system that can be audited for waiting times
- Appropriate referrals to specialist colleagues will be documented and GP informed of any transfer of care.
- All paediatric, adolescent and adult patients with congenital anomalies of the genital tract will be referred to a specialist centre.

Discharge criteria and planning

A discharge plan will be prepared with the patient offering support and facilities required for providing care at home. Post-surgical vaginal dilation supervision will be offered if required.

Patient-Centred Services

In all centres there will be a focus on patient centred services which consider the health and

well being needs of the patient with a complex congenital gynaecological anomaly. The importance of the family and close friends is acknowledged and supported. Services will include;

- Direct access (phone and e mail) to Clinical Nurse Specialist for clinical and treatment queries
- Transition facilitator – this may be a nurse or psychologist with specific responsibility for ensure smooth transition of adolescent patients
- Direct line/e mail contact to service administration for appointment/admission queries
- Provision of accurate and age-appropriate written information about the conditions and treatment options

Close links will be established with the well organised and active patient peer support groups in this area. Literature from support groups should be available in clinic and offered to patients who may wish to contact them. Support groups should be involved in service planning and feedback on services The major groups include;

- Androgen Insensitivity Syndrome Support Group (www.aissg.org)
- Congenital Adrenal Hyperplasia Support Group (www.livingwithcah.com)
- DSD Families (www.dsdfamilies.org)
- Turner Syndrome Support Group (www.tss.org.uk)

Other smaller groups exist and also have an important role to play.

Efforts will also be made to engage with and offer support to patients who elect not to be part of a support group but may have important input regarding service development

3.3 Population covered

The service outlined in this specification is for patients ordinarily resident in England*; or otherwise the commissioning responsibility of the NHS in England (as defined in Who Pays?: Establishing the responsible commissioner and other Department of Health guidance relating to patients entitled to NHS care or exempt from charges).

* - Note: for the purposes of commissioning health services, this EXCLUDES patients who, whilst resident in England, are registered with a GP Practice in Wales, but INCLUDES patients resident in Wales who are registered with a GP Practice in England.

Specifically this service is for adolescent girls and adult women with congenital anomalies of the genital tract requiring specialised intervention and management as outlined within this specification.

Currently up to 10 units in England offer this multidisciplinary service. Accurate incidence figures are not available but it is estimated this affects 1 in 2-3000 women.

3.4 Any acceptance and exclusion criteria and thresholds

This service does not accept patients with a diagnosis of gender dysphoria/transgender.

3.5 Interdependencies with other services/providers

i) Co-located Services

The multi-disciplinary team will include:

- Gynaecologist (including advanced minimal access surgery)
- Psychologist
- Specialist nurse
- Endocrinologist
- Urologist
- Radiologist
- Paediatric services should usually be co-located but in some areas may be in close proximity depending on hospital configuration

11) Interdependent Services

- Colorectal
- Biochemist
- Geneticist
- Plastic surgery, as required
- Paediatric endocrinology
- Paediatric urology/surgery
- Paediatric psychology

Related Services

- Primary Care
- Local Mental Health Services (Adolescent and Adult)

iv) Data Submission

- Number of new referrals per year
- Number of follow up appointments per year
 - Number of patients referred for vaginal dilation
 - Number of open surgical procedures
 - Number of laparoscopic surgical procedures

4. Applicable Service Standards

4.1 Applicable national standards e.g. NICE – See Appendix 3

STANDARDS OF CARE

- All patients must be under the care of an experienced multidisciplinary team
- Expert psychological input targeting emotional and social adjustment is crucial. Patients are likely to require interventions to cope with an atypical karyotype, the emotional challenges of infertility, complications related to sexual function, orientation and identity, communication barriers within the family, with friends and partners and problematic transitions.
- A supervised course of vaginal dilation must be the first line treatment for vaginal agenesis. Appropriate surgical treatments must be available for patients who fail or are unsuitable for dilation.
- Surgery must be performed by gynaecologists experienced in these procedures and where appropriate with input from other specialist surgical services such as urology, plastics and colorectal.
- Specific procedures must only be performed laparoscopically. These include gonadectomy, excision of uterine/mullerian remnant, uterovaginal anastomosis, Vecchietti procedure, Davidov procedure and some high transverse vaginal septums.
- Units must have close links with paediatric services and a specific focus on transition for adolescents.
- Specialised diagnostic support including genetics, biochemistry and imaging
- Fertility input may be required in some conditions for example IVF and ovum donation in POF, Turner's and Swyer syndromes as well as surrogacy in mullerian agenesis.
- Centres managing patient with complex congenital anomalies have a duty to:
 - a. Audit surgical and non-surgical outcomes
 - b. Build links with patient peer support groups
 - c. Participate in clinical networks mapped by BritSPAG
 - d. Contribute to the body of knowledge on The Service by publishing original research as well as commentaries and reviews
 - e. Contribute to the education of Specialist Registrars and other health professionals

Potential opportunities for future development

- National DSD Clinical Network
- Establish national DSD network for centres managing DSD. This could be in the forums of regular video conferencing to allow clinical collaboration with information sharing on complex or unusual cases as well as strategic work e.g. guidelines for best practice.
- Develop "Patient Navigator" Concept
Patient navigators are trained, culturally sensitive health care workers who provide support

and guidance for patients and families through the complexity of a DSD diagnosis. They are increasingly available in cancer services and have not as yet been used in a DSD setting although the complexity of the diagnosis would make this a worthwhile development

See Appendix 3 for references

DRAFT FOR PUBLIC CONSULTATION

Appendix 1

Quality standards specific to the service using the following template:

Quality Requirement	Threshold	Method of Measurement	Consequence of breach
Domain 1: Preventing people dying prematurely			
1. Gonadectomy in XY girls and women to prevent gonadal cancer	0% (All girls and women should be offered appropriately timed gonadectomy).	New cases of gonadal cancer as percentage of XY women	Audit to evaluate reasons why gonadectomy not performed.
1. Adequate hormone replacement therapy (HRT) post-gonadectomy and in ovarian failure	0% (All girls and women should be offered HRT to prevent osteoporosis and cardiac disease	Proportion of patients stabilised on HRT	Audit to evaluate reasons for patients not on HRT e.g compliance and change in practice where feasible
Domain 2: Enhancing the quality of life of people with long-term conditions			
1. Vaginal creation for vaginal agenesis, stenosis or cloacal anomaly by vaginal dilation	80%	Percentage of girls or women sexually active at completion of dilator programme	Audit of success rates following dilation
2. Vaginal reconstruction for those where dilation has failed or is not appropriate	>90% - all women should be offered surgery	Percentage of girls or women sexually active after surgery	Audit of success rates following surgery. Include comparison of different available reconstructive procedures.
Domain 3: Helping people to recover from episodes of ill-health or following injury			
1. Psychological support for disclosure of XY karyotype, absent uterus and/or vagina, lack of fertility	100% All adolescent girls and women must be offered psychology.	Proportion of patients seeing psychologist	Audit to evaluate why patients are not seeing psychologist
Domain 4: Ensuring that people have a positive experience of care			
1. Patient satisfaction with	100% of patients offered survey	Patient satisfaction scores	Audit proportion of patients surveyed and reasons for

Quality Requirement	Threshold	Method of Measurement	Consequence of breach
<p>service – annual survey</p> <p>2. Named key worker for transition from paediatric to adolescent and adult services</p>	100%	Percentage of adolescent patients with named key worker	<p>dissatisfaction</p> <p>Audit to evaluate reasons why no transition worker</p>
Domain 5: Treating and caring for people in a safe environment and protecting them from avoidable harm			
1. Patients with vaginal agenesis treated with vaginal dilation as first line	100% patients with vaginal agenesis	Proportion of patients referred to clinical nurse specialist dilation clinic	Audit of patients declining or failing dilation therapy.
2. Patients treated laparoscopically where possible and appropriate	100% of patients requiring specific surgical intervention	Proportion of suitable patients receiving laparoscopic procedures	Audit morbidity, inpatient length of stay and cost of surgical vaginoplasty
3. Patients treated surgically in a centre commissioned for and compliant with national program of care	100% All patients requiring surgical intervention	Number of patients treated surgically at other units prior to referral	Audit of inappropriate or inadequate surgery
4. Compliance with BritSPAG Guidelines for Complex PAG	100%	Annual BritSPAG Survey of Units	Audit to evaluate reasons for non-compliance and recommend remedial action

APPENDIX 2

Service Pathway – Complex Gynaecology Congenital Gynaecological Anomalies		
Referral Pathway type/source	Service Delivery	
<p>Secondary New referral (gynaecology or paediatrics) or from specialist paediatrics or new adolescent transition from specialised paediatric services</p>	<p>Primary GP</p>	<p>Tertiary From specialist paediatric services for transition</p>
<p>Outpatient Appointment (Detail? Assessment, MDT, Single Clinician etc)</p>	<p>Multi-disciplinary Outpatient Clinic Core team Gynaecology, Psychology, Specialised Nursing, Access to Biochemistry, Genetics Imaging,</p>	
<p>Investigations/procedures needed</p>	<p>Bloods; Biochemistry, Karyotype and Genetic Imaging; Ultrasound and MRI, Bone mineral density Examination under anaesthetic +/- Cystoscopy and Vaginocopy</p>	
<p>Treatment Strategy (please provide detail)</p>	<p>Inpatient Must include advanced minimal access surgery</p> <p>Procedures include; Gonadectomy, Vaginoplasty (all types), Clitoral reduction, Excision of septum, Excision of obstructed mullerian remnant, Uterovaginal anastomosis</p>	<p>Outpatient</p> <p>Psychology Vaginal Dilation Therapy Induction of puberty</p>
<p>Follow up (detail)</p>	<p>Post-surgical (dilation). 1, 6 and 12 weeks follow-up. Post-surgical (no dilation) 6 weeks follow up Dilation programme (no surgery). 2 weekly nurse led follow up for up to 6 months. Psychology and psychosexual input as required. Post-gonadectomy - oestrogen replacement, 6 weeks</p>	
<p>Further investigations/treatments required (detail)</p>	<p>Post-gonadectomy - oestrogen replacement annual Psychology and psychosexual. Fertility referral Chronic Disease Surveillance</p>	
<p>Second treatment episode if needed (detail)</p>	<p>Complications such as vaginal stenosis require dilation+/- repeat vaginoplasty Failed primary vaginal dilation treatment (up to 20%) process to reconstructive surgery Psychology and psychosexual</p>	

APPENDIX 3

References

1. Consensus statement on management of intersex disorders Hughes et al Arch.Dis.Child July 91(7) 554-63
2. UK Guidance on the initial evaluation of an infant or an adolescent with a suspected disorder of sex development. Society for Endocrinology Clinical Guideline Article. Clin. Endo 2011 75:12-26
3. Clinical Standards for Service Planning in Paediatric and Adolescent Gynaecology. British Society for Paediatric and Adolescent Gynaecology. January 2011 www.britspag.org
4. Congenital Adrenal Hyperplasia in the Adult. British Society for Endocrinology Guidelines 2013
5. American College of Obstetricians and Gynaecologists. Mullerian agenesis: diagnosis, management and treatment. Committee opinion Number 562 May 2013

APPENDIX 4

Incidence of conditions

The table below gives incidences of some of the conditions covered by this service specification. The current female population of England is approximately 25 Million. Just taking the above conditions gives a minimum of 0.5 to 1 per 1,000 population i.e potentially 25,000 cases.

Condition	Incidence	Reference
Mayer-Rokitansky-Kuster-Hauser Syndrome	1 in 5000	Attomaki et al Fert.Steril. 2001
Congenital Adrenal Hyperplasia	1 in 30,000 female	Pang SY et al. Paediatrics 1988
Complete Androgen Insensitivity Syndrome	1 in 40,000	Boehmer AL et al. J.Endocrin.Metab 2001
Turner Syndrome	1 in 3,000	Gravholt, S. Juul, et al BMJ. 1996
Complex Mullerian Anomalies (transverse septae, obstructed hemivagina etc)	Approx. 1 in 5,000	No recent medical literature

APPENDIX 5

ANNEX 1 TO SERVICE SPECIFICATION:

PROVISION OF SERVICES TO CHILDREN

Aims and objectives of service

This specification annex applies to all children's services and outlines generic standards and outcomes that would be fundamental to all services.

The generic aspects of care:

The Care of Children in Hospital (Health Service Circular 1998/238) requires that:

- Children are admitted to hospital only if the care they require cannot be as well provided at home, in a day clinic or on a day basis in hospital.
- Children requiring admission to hospital are provided with a high standard of medical, nursing and therapeutic care to facilitate speedy recovery and minimize complications and mortality.
- Families with children have easy access to hospital facilities for children without needing to travel significantly further than to other similar amenities.
- Children are discharged from hospital as soon as socially and clinically appropriate and full support provided for subsequent home or day care.
- Good child health care is shared with parents/carers and they are closely involved in the care of their children at all times unless, exceptionally, this is not in the best interest of the child. Accommodation is provided for them to remain with their children overnight if they so wish.

Service description/care pathway

All paediatric specialised services have a component of primary, secondary, tertiary and even quaternary elements.

The efficient and effective delivery of services requires children to receive their care as close to home as possible dependent on the phase of their disease.

Services should therefore be organised and delivered through "integrated pathways of care" (National Service Framework for children, young people and maternity services (Department of Health & Department for Education and Skills, London 2004)

Interdependencies with other services

All services will comply with *Commissioning Safe and Sustainable Specialised Paediatric Services: A Framework of Critical Inter-Dependencies* – Department of Health (DH)

Imaging

All services will be supported by a 3 tier imaging network ('Delivering quality imaging services for children' DH 13732 March 2010). Within the network:

- It will be clearly defined which imaging test or interventional procedure can be performed and reported at each site
- Robust procedures will be in place for image transfer for review by a specialist radiologist, these will be supported by appropriate contractual and information governance arrangements
- Robust arrangements will be in place for patient transfer if more complex imaging or intervention is required
- Common standards, protocols and governance procedures will exist throughout the network.
- All radiologists, and radiographers will have appropriate training, supervision and access to continuing professional development
- All equipment will be optimised for paediatric use and use specific paediatric software.

Specialist Paediatric Anaesthesia

Wherever and whenever children undergo anaesthesia and surgery, their particular needs must be recognised and they should be managed in separate facilities, and looked after by staff with appropriate experience and training.¹ All UK anaesthetists undergo training which provides them with the competencies to care for older babies and children with relatively straightforward surgical conditions and without major co-morbidity. However those working in specialist centres must have undergone additional (specialist) training² and should maintain the competencies so acquired³ *. These competencies include the care of very young/premature babies, the care of babies and children undergoing complex surgery and/or those with major/complex co-morbidity (including those already requiring intensive care support).

As well as providing essential co-dependent service for surgery specialist anaesthesia and sedation services may be required to facilitate radiological procedures and interventions (for example MRI scans and percutaneous nephrostomy) and medical interventions (for example joint injection and intrathecal chemotherapy), and for assistance with vascular access in babies and children with complex needs such as intravenous feeding.

Specialist acute pain services for babies and children are organised within existing departments of paediatric anaesthesia and include the provision of agreed (hospital wide) guidance for acute pain, the safe administration of complex analgesia regimes including epidural analgesia, and the daily input of specialist anaesthetists and acute pain nurses with expertise in paediatrics.

*The Safe and Sustainable reviews of paediatric cardiac and neuro- sciences in England have noted the need for additional training and maintenance of competencies by specialist anaesthetists in both fields of practice.

References

1. Guidelines for the Provision of Anaesthetic Services (GPAS) Paediatric anaesthetic services. RCoA 2010 www.rcoa.ac.uk
2. Certificates of Completion of Training (CCT) in Anaesthesia 2010
3. Continuing Professional Development (CPD) matrix level 3

Specialised Child and Adolescent Mental Health Services (CAMHS)

The age profile of children and young people admitted to specialised CAMHS day/in-patient settings is different to the age profile for paediatric units in that it is predominantly adolescents who are admitted to specialised CAMHS in-patient settings, including over-16s. The average length of stay is longer for admissions to mental health units. Children and young people in specialised CAMHS day/in-patient settings generally participate in a structured programme of education and therapeutic activities during their admission.

Taking account of the differences in patient profiles the principles and standards set out in this specification apply with modifications to the recommendations regarding the following:

- Facilities and environment – essential Quality Network for In-patient CAMHS (QNIC) standards should apply (<http://www.rcpsych.ac.uk/quality/quality accreditationaudit/qnic1.aspx>)
- Staffing profiles and training - essential QNIC standards should apply.
- The child/ young person's family are allowed to visit at any time of day taking account of the child / young persons need to participate in therapeutic activities and education as well as any safeguarding concerns.
- Children and young people are offered appropriate education from the point of admission.
- Parents/carers are involved in the child/young persons care except where this is not in the best interests of the child / young person and in the case of young people who have the capacity to make their own decisions is subject to their consent.
- Parents/carers who wish to stay overnight are provided with accessible accommodation unless there are safeguarding concerns or this is not in the best interests of the child/ young person.

Applicable national standards e.g. NICE, Royal College

Children and young people must receive care, treatment and support by staff registered by the Nursing and Midwifery Council on the parts of their register that permit a nurse to work with children (Outcome 14h Essential Standards of Quality and Safety, Care Quality Commission, London 2010)

- There must be at least two Registered Children's Nurses (RCNs) on duty 24 hours a day in all hospital children's departments and wards.
- There must be an Registered Children's Nurse available 24 hours a day to advise on the nursing of children in other departments (this post is included in the staff establishment of 2RCNs in total).

Accommodation, facilities and staffing must be appropriate to the needs of children and separate from those provided for adults. All facilities for children and young people must comply with the Hospital Build Notes HBN 23 Hospital Accommodation for Children and Young People NHS Estates, The Stationary Office 2004.

All staff who work with children and young people must be appropriately trained to provide care, treatment and support for children, including Children's Workforce Development Council Induction standards (Outcome 14b Essential Standards of Quality and Safety, Care Quality Commission, London 2010).

Each hospital who admits inpatients must have appropriate medical cover at all times taking account of guidance from relevant expert or professional bodies (National Minimum Standards for Providers of Independent Healthcare, Department of Health, London 2002). "Facing the Future" Standards, Royal College of Paediatrics and Child Health.

Staff must carry out sufficient levels of activity to maintain their competence in caring for children and young people, including in relation to specific anaesthetic and surgical procedures for children, taking account of guidance from relevant expert or professional bodies (Outcome 14g Essential Standards of Quality and Safety, Care Quality Commission, London 2010).

Providers must have systems in place to gain and review consent from people who use services, and act on them (Outcome 2a Essential Standards of Quality and Safety, Care Quality Commission, London 2010). These must include specific arrangements for seeking valid consent from children while respecting their human rights and confidentiality and ensure that where the person using the service lacks capacity, best interest meetings are held with people who know and understand the person using the service. Staff should be able to show that they know how to take appropriate consent from children, young people and those with learning disabilities (Outcome 2b) (Seeking Consent: working with children Department of Health, London 2001).

Children and young people must only receive a service from a provider who takes steps to prevent abuse and does not tolerate any abusive practice should it occur (Outcome 7 Essential Standards of Quality and Safety, Care Quality Commission, London 2010 defines the standards and evidence required from providers in this regard). Providers minimise the risk and likelihood of abuse occurring by:

- Ensuring that staff and people who use services understand the aspects of the safeguarding processes that are relevant to them
- Ensuring that staff understand the signs of abuse and raise this with the right person when those signs are noticed.
- Ensuring that people who use services are aware of how to raise concerns of abuse.
- Having effective means to monitor and review incidents, concerns and complaints that have the potential to become an abuse or safeguarding concern.
- Having effective means of receiving and acting upon feedback from people who use services and any other person.
- Taking action immediately to ensure that any abuse identified is stopped
- and suspected abuse is addressed by:
 - a. Having clear procedures followed in practice, monitored and reviewed that take account of relevant legislation and guidance for the management of alleged abuse

- b. Separating the alleged abuser from the person who uses services and others who may be at risk or managing the risk by removing the opportunity for abuse to occur, where this is within the control of the provider
 - c. Reporting the alleged abuse to the appropriate authority
 - d. Reviewing the person's plan of care to ensure that they are properly supported following the alleged abuse incident.
- Using information from safeguarding concerns to identify non-compliance, or any risk of non-compliance, with the regulations and to decide what will be done to return to compliance.
 - Working collaboratively with other services, teams, individuals and agencies in relation to all safeguarding matters and has safeguarding policies that link with local authority policies.
 - Participates in local safeguarding children boards where required and understand their responsibilities and the responsibilities of others in line with the Children Act 2004.
 - Having clear procedures followed in practice, monitored and reviewed in place about the use of restraint and safeguarding.
 - Taking into account relevant guidance set out in the Care Quality Commission's Schedule of Applicable Publications
 - Ensuring that those working with children must wait for a full CRB disclosure before starting work.
 - Training and supervising staff in safeguarding to ensure they can demonstrate the competences listed in Outcome 7E of the Essential Standards of Quality and Safety, Care Quality Commission, London 2010

All children and young people who use services must be:

- Fully informed of their care, treatment and support.
- Able to take part in decision making to the fullest extent that is possible.
- Asked if they agree for their parents or guardians to be involved in decisions they need to make.

(Outcome 4I *Essential Standards of Quality and Safety*, Care Quality Commission, London 2010)

Key Service Outcomes

Evidence is increasing that implementation of the national Quality Criteria for Young People Friendly Services (Department of Health, London 2011) have the potential to greatly improve patient experience, leading to better health outcomes for young people and increasing socially responsible life-long use of the NHS.

Implementation is also expected to contribute to improvements in health inequalities and public health outcomes e.g. reduced teenage pregnancy and STIs, and increased smoking cessation. All providers delivering services to young people should be implementing the good practice guidance which delivers compliance with the quality criteria.

Poorly planned transition from young people's to adult-oriented health services can be associated with increased risk of non adherence to treatment and loss to follow-up, which can have serious consequences. There are measurable adverse consequences in terms

of morbidity and mortality as well as in social and educational outcomes. When children and young people who use paediatric services are moving to access adult services (for example, during transition for those with long term conditions), these should be organised so that:

- All those involved in the care, treatment and support cooperate with the planning and provision to ensure that the services provided continue to be appropriate to the age and needs of the person who uses services.

The National Minimum Standards for Providers of Independent Healthcare, (Department of Health, London 2002) require the following standards:

- **A16.1** Children are seen in a separate out-patient area, or where the hospital does not have a separate outpatient area for children, they are seen promptly.
- **A16.3** Toys and/or books suitable to the child's age are provided.
- **A16.8** There are segregated areas for the reception of children and adolescents into theatre and for recovery, to screen the children and adolescents from adult patients; the segregated areas contain all necessary equipment for the care of children.
- **A16.9** A parent is to be actively encouraged to stay at all times, with accommodation made available for the adult in the child's room or close by.
- **A16.10** The child's family is allowed to visit him/her at any time of the day, except where safeguarding procedures do not allow this
- **A16.13** When a child is in hospital for more than five days, play is managed and supervised by a qualified Hospital Play Specialist.
- **A16.14** Children are required to receive education when in hospital for more than five days; the Local Education Authority has an obligation to meet this need and are contacted if necessary.
- **A18.10** There are written procedures for the assessment of pain in children and the provision of appropriate control.

All hospital settings should meet the Standards for the Care of Critically Ill Children (Paediatric Intensive Care Society, London 2010).

There should be age specific arrangements for meeting Regulation 14 of the Health and Social Care Act 2008 (Regulated Activities) Regulations 2010. These require:

- A choice of suitable and nutritious food and hydration, in sufficient quantities to meet service users' needs
- Food and hydration that meet any reasonable requirements arising from a service user's religious or cultural background
- Support, where necessary, for the purposes of enabling service users to eat and drink sufficient amounts for their needs
- For the purposes of this regulation, "food and hydration" includes, where applicable, parenteral nutrition and the administration of dietary supplements where prescribed
- Providers must have access to facilities for infant feeding, including facilities to support breastfeeding (Outcome 5E, of the Essential Standards of Quality and Safety, Care Quality Commission, London 2010)

All paediatric patients should have access to appropriately trained paediatric trained dieticians, physiotherapists, occupational therapists, speech and language therapy, psychology, social work and CAMHS services within nationally defined access standards.

All children and young people should have access to a professional who can undertake an assessment using the Common Assessment Framework and access support from social care, housing, education and other agencies as appropriate.

All registered providers must ensure safe use and management of medicines, by means of the making of appropriate arrangements for the obtaining, recording, handling, using, safe keeping, dispensing, safe administration and disposal of medicines (Outcome 9 Essential Standards of Quality and Safety, Care Quality Commission, London 2010). For children, these should include specific arrangements that:

- They are supported to have a health action plan
- Facilities meet the appropriate requirements of the Disability Discrimination Act 1995

They meet the standards set out in Transition: getting it right for young people. Improving the transition of young people with long-term conditions from children's to adult health services. Department of Health, 2006, London.