The Clinical Nurse Specialist role in the DSD service in London, United Kingdom

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Conflict of Interest Disclosure

- Paediatric Endocrine Nurse Advisory boards – UK
  - Ipsen Ltd
  - Ferring Pharmaceuticals
  - Sandoz
- Invited lectures at Pharmaceutical company meetings
  - Ferring pharmaceuticals
  - Merck Serono
  - Novo Nordisk
  - Pfizer
  - Sandoz
- Winner of Ipsen BSPED Paediatric Endocrine Nurse Award 2014

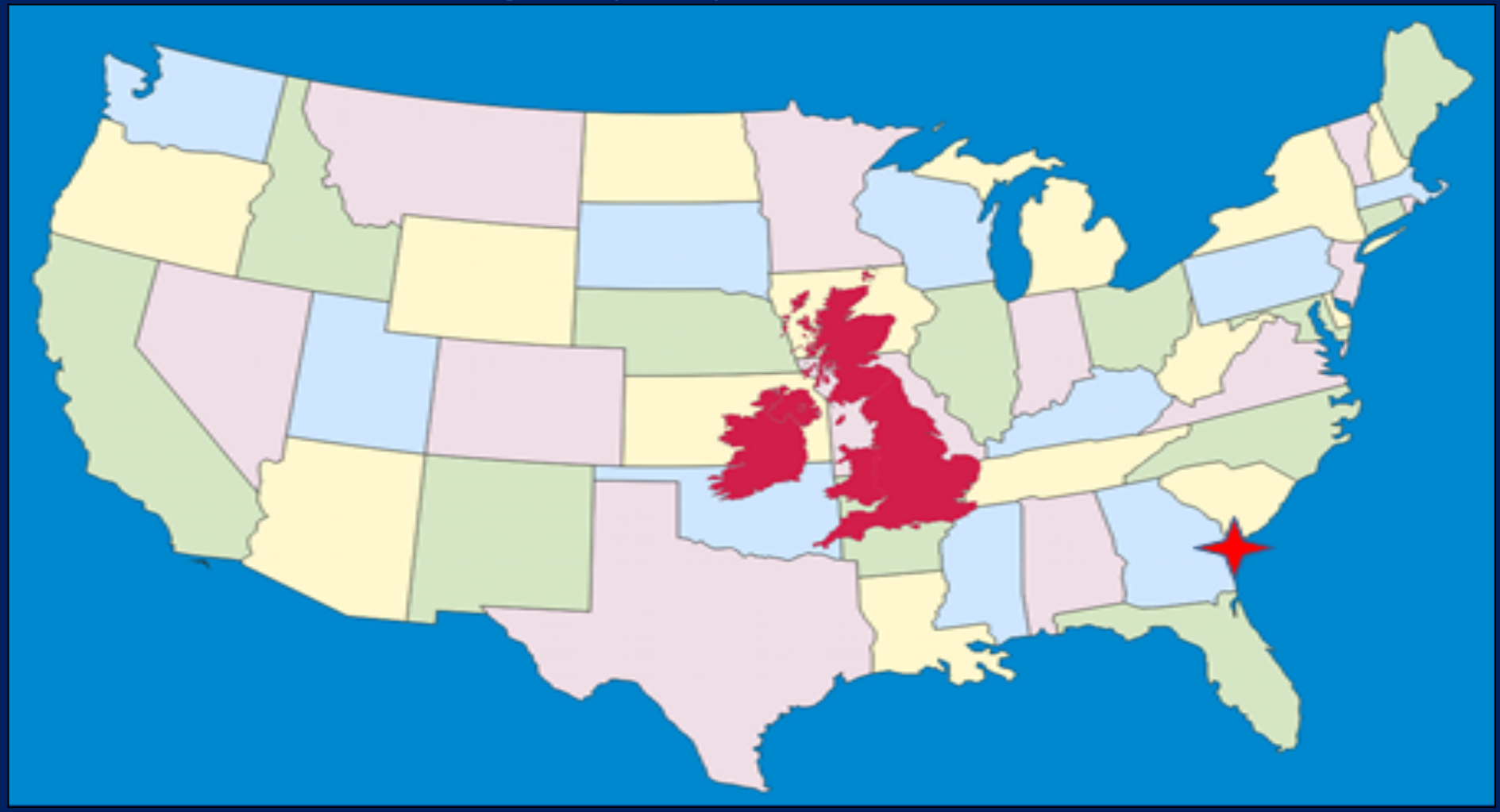
Introduction

- Objectives
- Geography
- The British National Health Service system
- What is a DSD?
- Our DSD team and service
- Referrals to GOSH
- The team role
- The CNS role
  - DSD
  - Adrenal
- Conclusion
- References

Objectives

- Describe the referral process for a baby with a DSD within the British National Health Service (NHS)
- Discuss the Great Ormond Street Hospital for children diagnostic flow chart
- Identify the role of the Clinical Nurse Specialist (CNS) in the DSD multi-disciplinary team

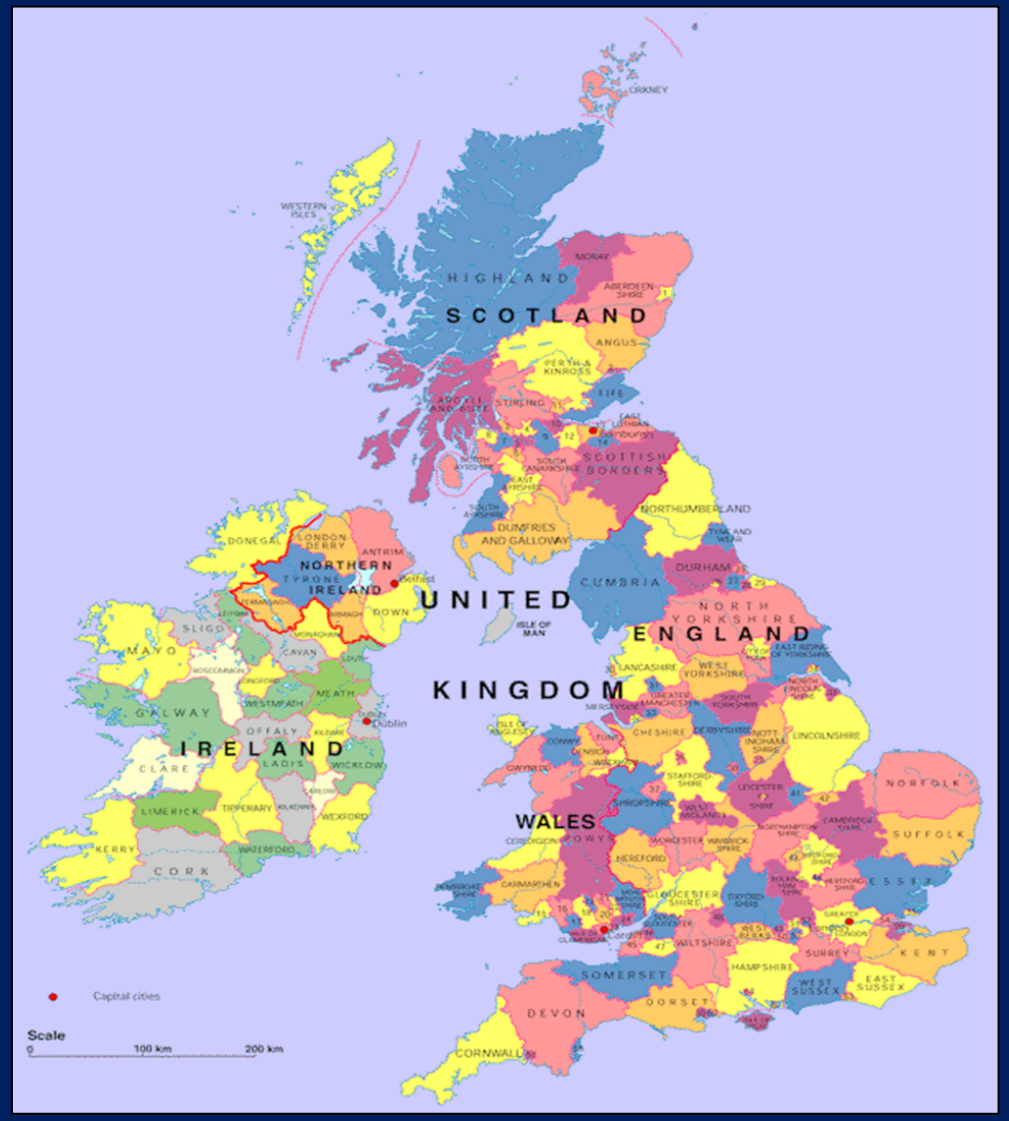
Geography and statistics

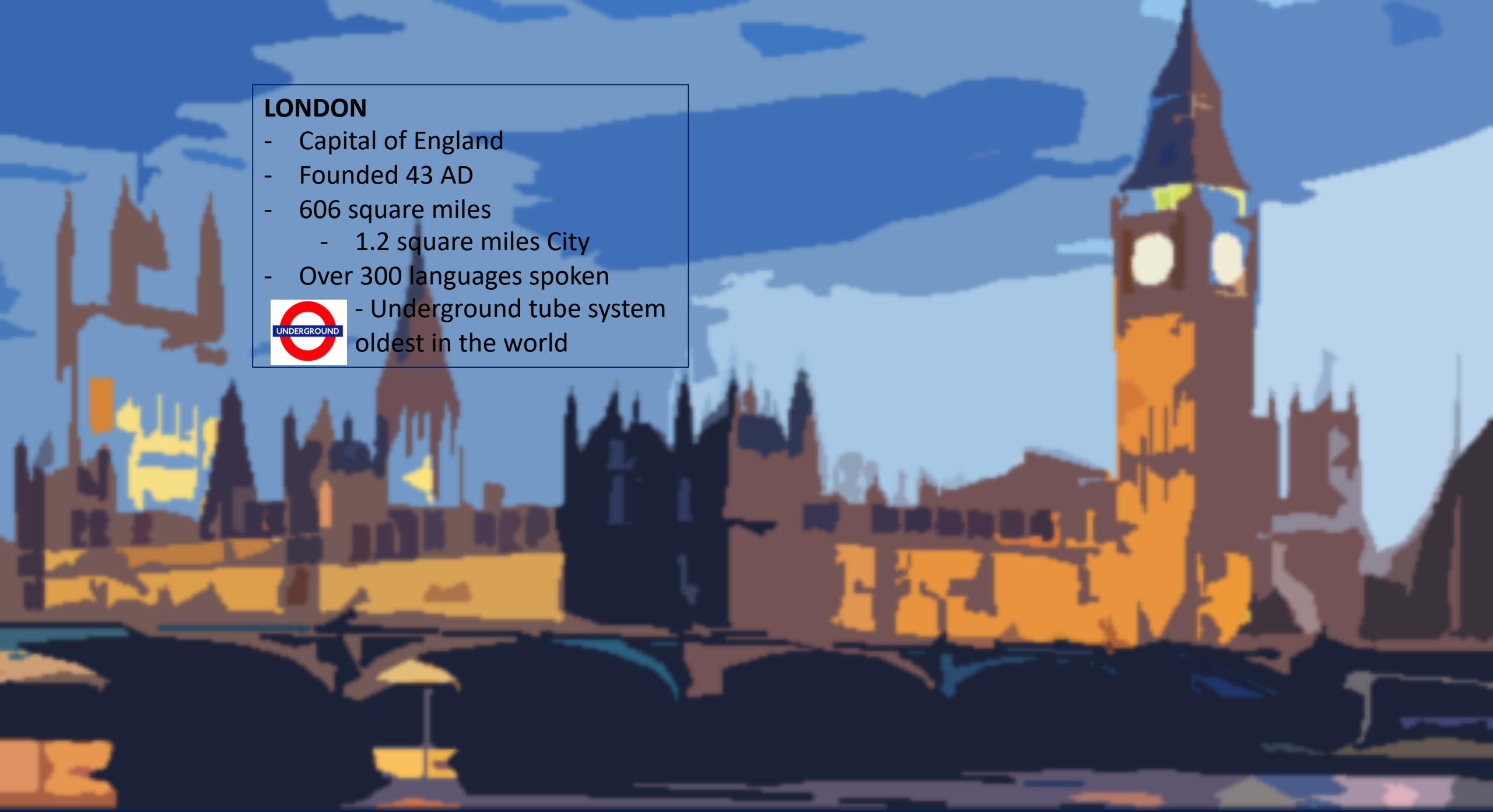


Relative sizes and population

- Most of England, Scotland and Wales covering most of Missouri and Iowa
  - Missouri: 6.1 million
  - Iowa: 3.1 million
- UK population: 64.1 million
- USA population: 318.9 million
- London population: 8.6 million
  - Los Angeles: 10 million

United Kingdom





LONDON

- Capital of England
- Founded 43 AD
- 606 square miles
  - 1.2 square miles City
- Over 300 languages spoken
- Underground tube system oldest in the world

Great Ormond Street Hospital for Children

- Opened in 1852
- Patroned
  - Queen Victoria → Princess Diana
  - Charles Dickens
  - JM Barrie – Peter Pan
- 387 patient beds
- > 50 clinical specialities
- > 240,000 patient visits per year
- 50% patients come from outside London
- Tertiary hospital
  - No ER

The British National Health System

- Formed in 1948 to provide free healthcare for all
  - Antenatal, maternity, postnatal, immunisations, child health, screening, ER
- Primary healthcare
  - General Practitioners, Practice Nurses, Health Visitors, School Nurses
  - Day to day healthcare
- Secondary healthcare
  - Provided by medical specialists – 'hospital care'
  - Referrals made by the patient's GP
- Tertiary healthcare
  - More highly specialised healthcare
  - Referral made by a secondary healthcare professional

What is a DSD?

- Congenital conditions in which development of chromosomal, gonadal or anatomic sex is atypical
- True genital ambiguity
  - 1 in 5000 / 1 in 4500 births
- Genital anomalies
  - 1 in 300 births

Classification of DSD

- 46,XY DSD (under virilised genetic male)
  - Disorders of testicular development
    - Ovotesticular DSD
  - Disorders of androgen synthesis / action
    - CAIS
  - Others
    - Hypospadias
- 46,XX DSD (over virilised genetic female)
  - Disorders of ovarian development
    - Ovotesticular DSD
  - Androgen excess
    - CAH

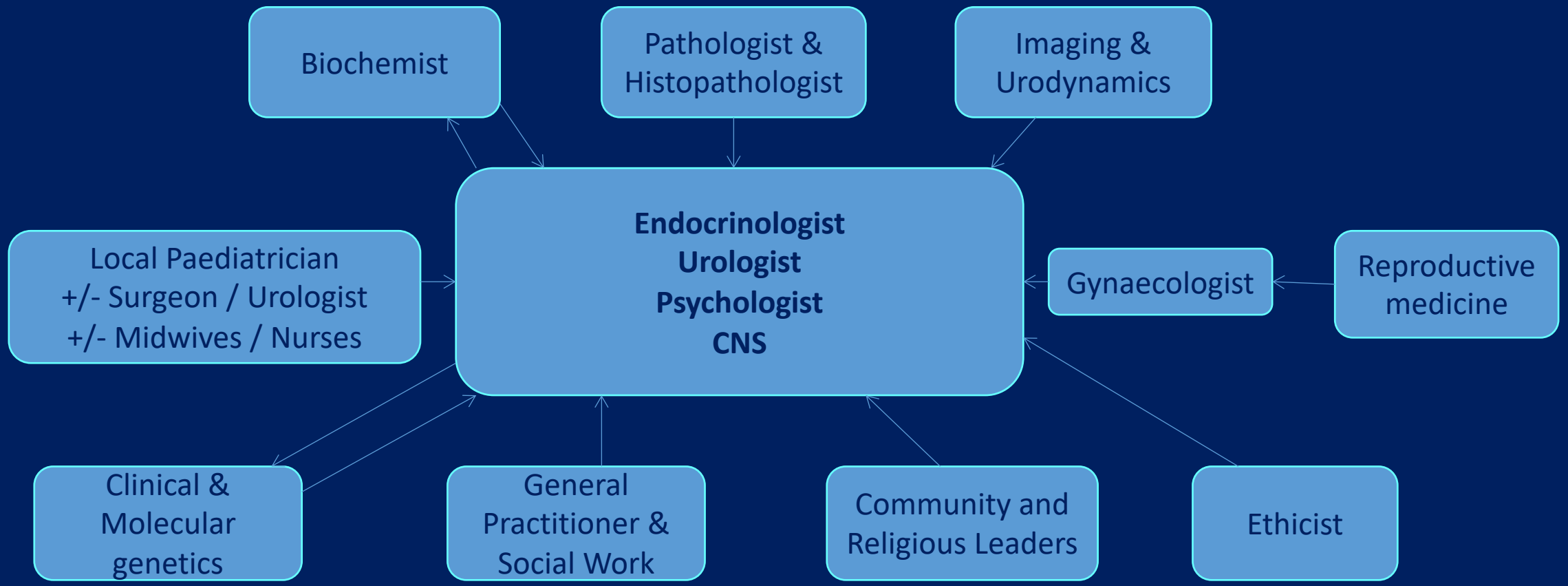
- Sex chromosome DSD (variable)
  - Turner's syndrome
  - Klinefelter's syndrome
  - Mixed gonadal dysgenesis

Sex Chromosome DSD	DSD 46,XY	DSD 46,XX
45,X (Turner Syndrome and variants)	Disorders of gonadal (testicular) development: 1) Complete gonadal dysgenesis (Swyer Syndrome) 2) Parcial gonadal dysgenesis 3) Gonadal regression 4) Ovotesticular DSD 5) CBX2 gene def. (ovaries + fem. ext. gen.)	Disorders of gonadal (ovarian) development: 1) Ovotesticular DSD 2) Testicular DSD (SRY^+, duplication of SOX9), 46,XX males. Def. gen 3) Gonadal dysgenesis
47,XXY (Klinefelter Syndrome and variants)	Disorders of androgen synthesis or action: 1) Androgen biosynthesis defects (17-hydroxylase, 5αRD2, StAR protein, 3β-HSD, 17β-HSD) 2) Defects in androgen actions (CAIS, PAIS) 3) Defects in LH receptor (Leydig cell hypoplasia) 4) Defects in AMH or AMH receptor (Persistence Müllerian ducts syndrome)	Androgen excess: 1) Fetal (Defects in 21-hydroxilase, or 11-hydroxylase) 2) Fetoplacental (deficiencia de aromatasa, POR [P450 oxidoreductasa]) 3) Maternal (luteoma, exogenous androgens, etc)
45,X/46,XY (mixed gonadal dysgenesis, ovotesticular DSD)	Other (cloacal extrophy, severe hypospadias)	Other (cloacal extrophy, vaginal atresia, other)
46,XX/46,XY (chimeric, ovotesticular DSD)		

Revised nomenclature: Chicago Consensus 2006

Previous	Revised
Intersex	Disorders of sex development (DSDs)
Male pseudohermaphrodite	
Undervirilization of an XY male	46,XY DSD
Undermasculinization of an XY male	
Female pseudohermaphrodite	
Overvirilization of an XX female	46,XX DSD
Masculinization of an XX female	
True hermaphrodite	Ovotesticular DSD
XX male or XX sex reversal	46,XX testicular DSD
XY sex reversal	46,XY complete gonadal dysgenesis

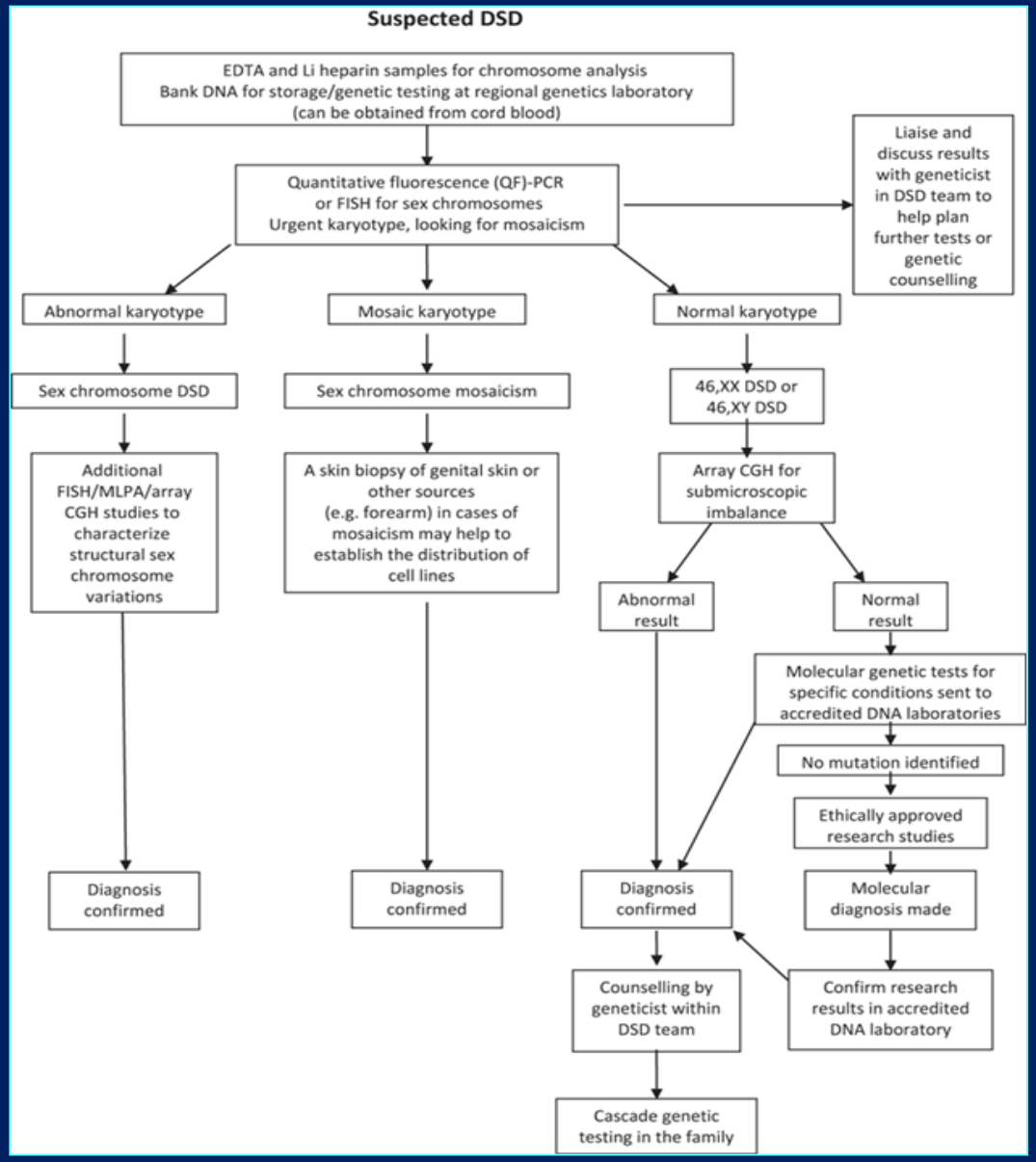
DSD MDT team at GOSH



Endocrine CNS team

- Congenital Hypothyroidism
- Congenital Hyperinsulinism
- Neuro Endocrine Oncology
- Hypopituitarism / Septo-Optic Dysplasia
- Adrenal / DSD

Clinical actions – UK approach *(Ahmed, 2011)*



Monthly MDT meeting

- Every 2nd Monday of the month
- All meet
  - Lead Consultant Endocrinologist with DSD interest chairs the meeting
  - Registrars (Residents) present new cases
    - Discuss previous cases
      - Attended outpatients clinic
      - Attended the endocrine day case unit (Kingfisher)
      - Had EUA (Woodpecker)
      - Had surgery (Squirrel)
  - CNS
    - Makes notes on planned outcomes and proposed interventions.
      - CNS team ipad
      - Emailed to relevant people with tasks to be done

CNS DSD MDT notes

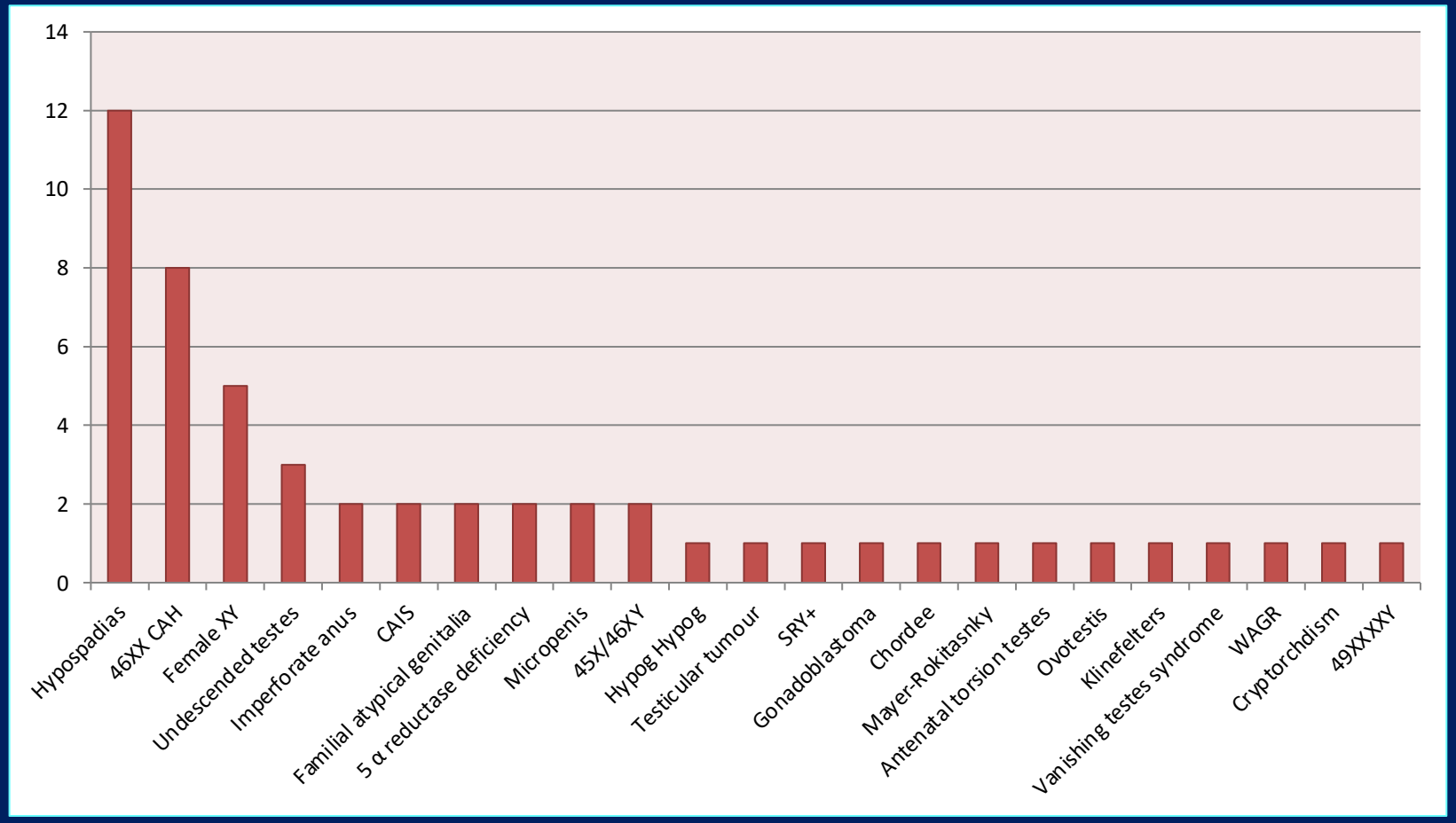
DSD MONDAY MDT ACTION PLAN

Date:

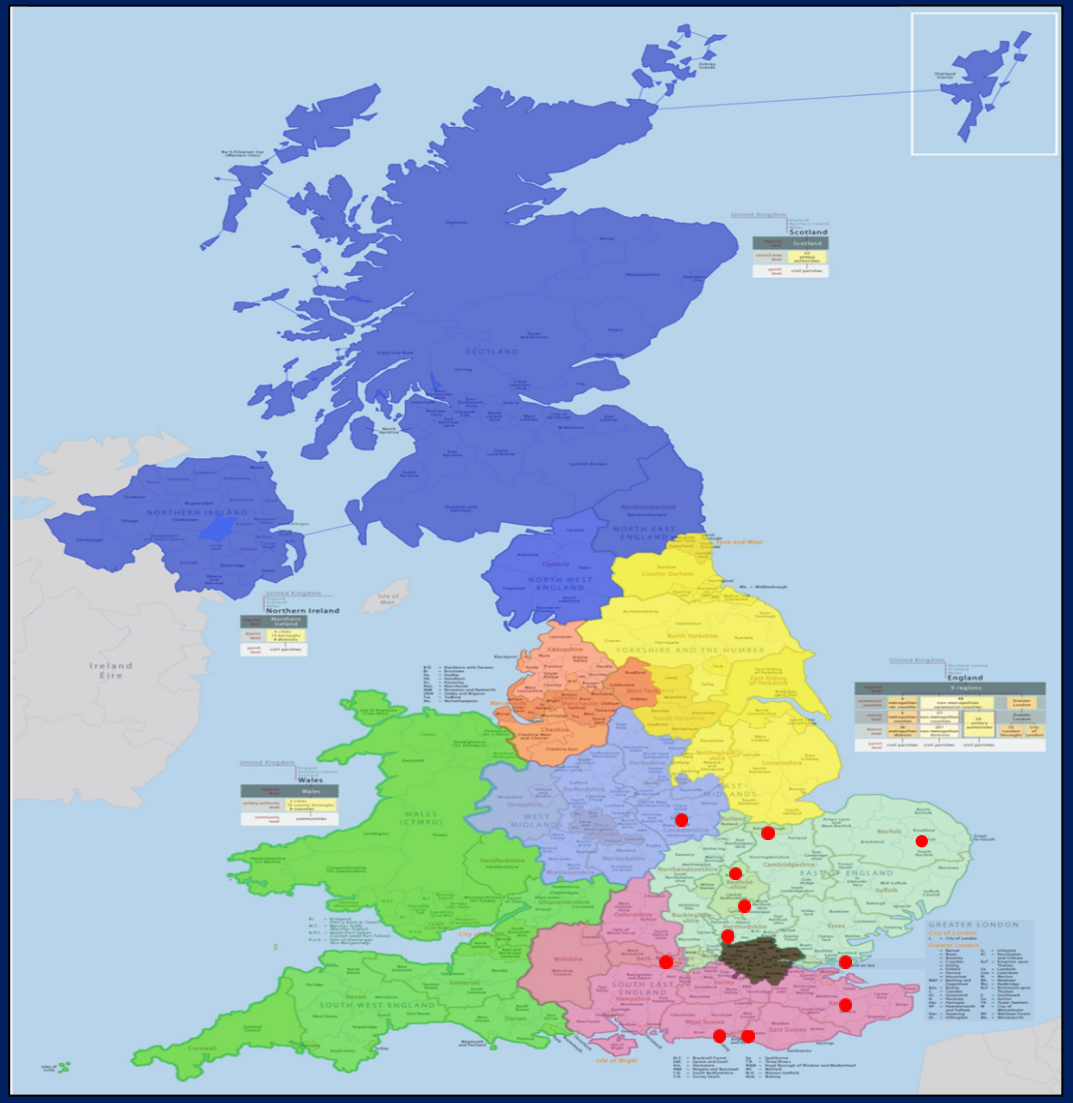
Name	Hosp No	Cons	Referring Hosp	Diagnosis	Plan	Action

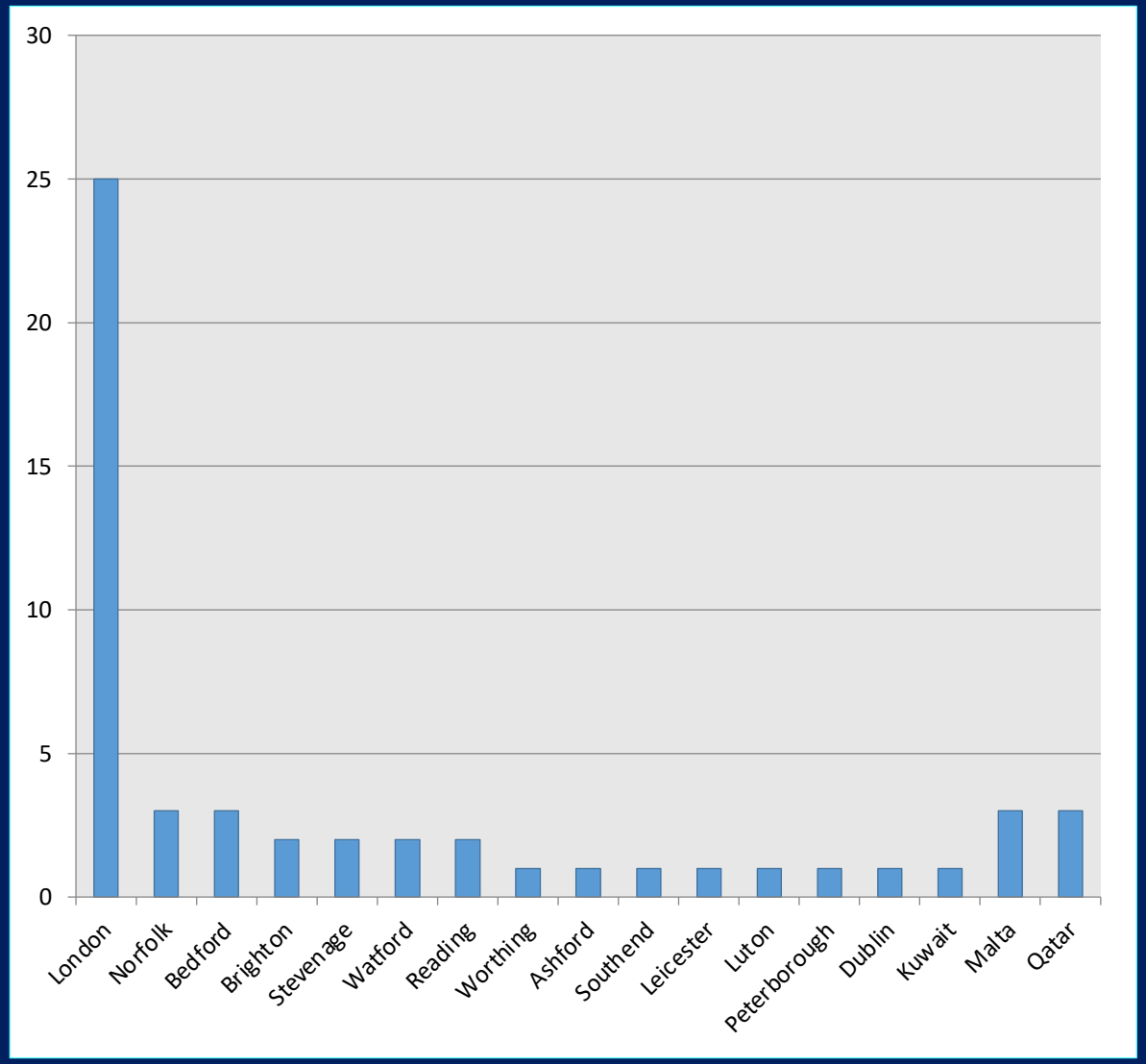
GOSH DSD diagnosis statistics - 2014

- 53 new referrals over one year
- Averaging 6 a month

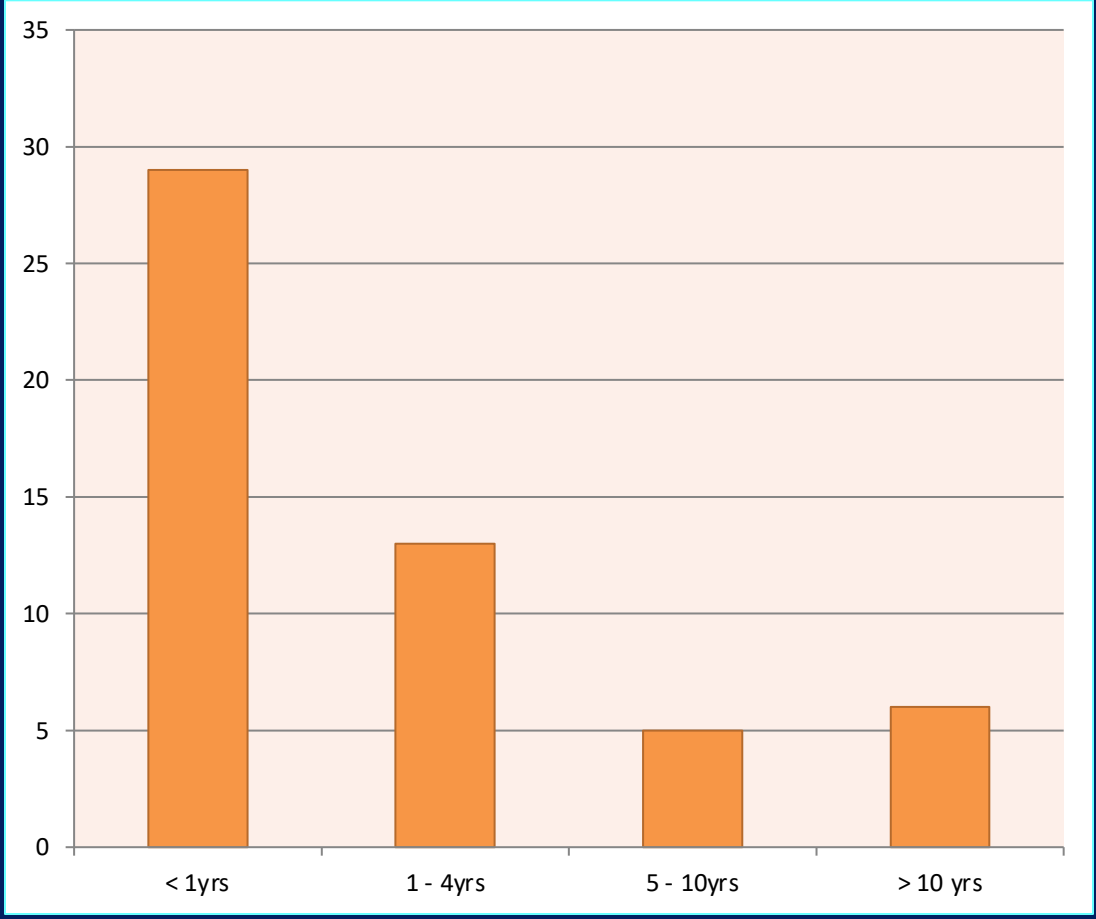


DSD referral areas



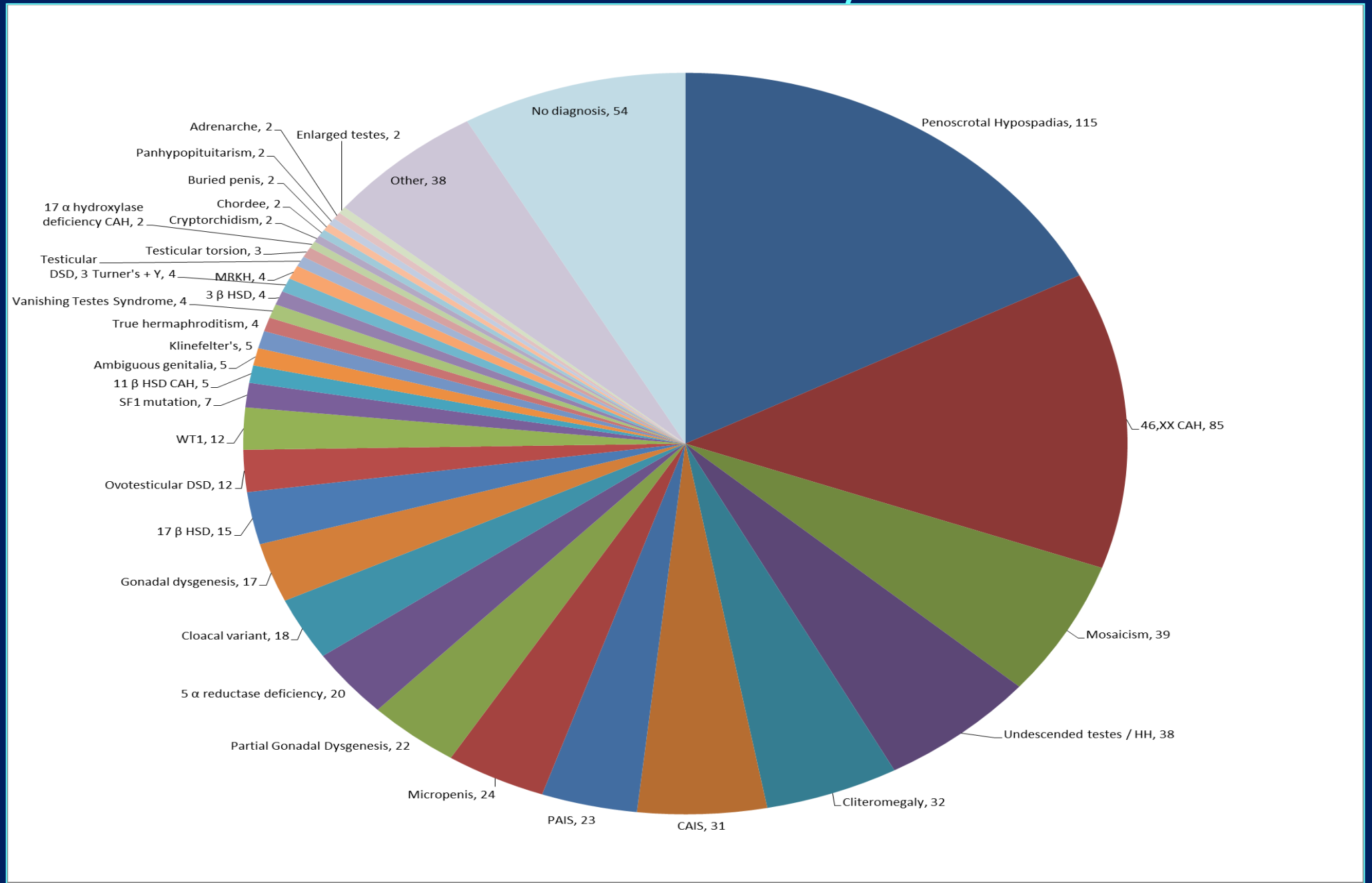


DSD ages of referral at GOSH



- Infants
  - Usually present with atypical genitalia
- Adolescents
  - Atypical sexual development
    - Micropenis
    - Cryptorchidism
    - Referrals from other centres
      - Previous hypospadias surgeries
      - Familial atypical genitalia
      - CAIS

GOSH DSD data over 21 years N= 657



Other – 38 single diagnoses

- Kallmann's syndrome
- IMAGE syndrome
- Labial fusion
- Accessory phallus
- Clitoral hypertrophy
- Absent vagina / ovary
- Atrophic right testes
- WAGR syndrome
- Tumours

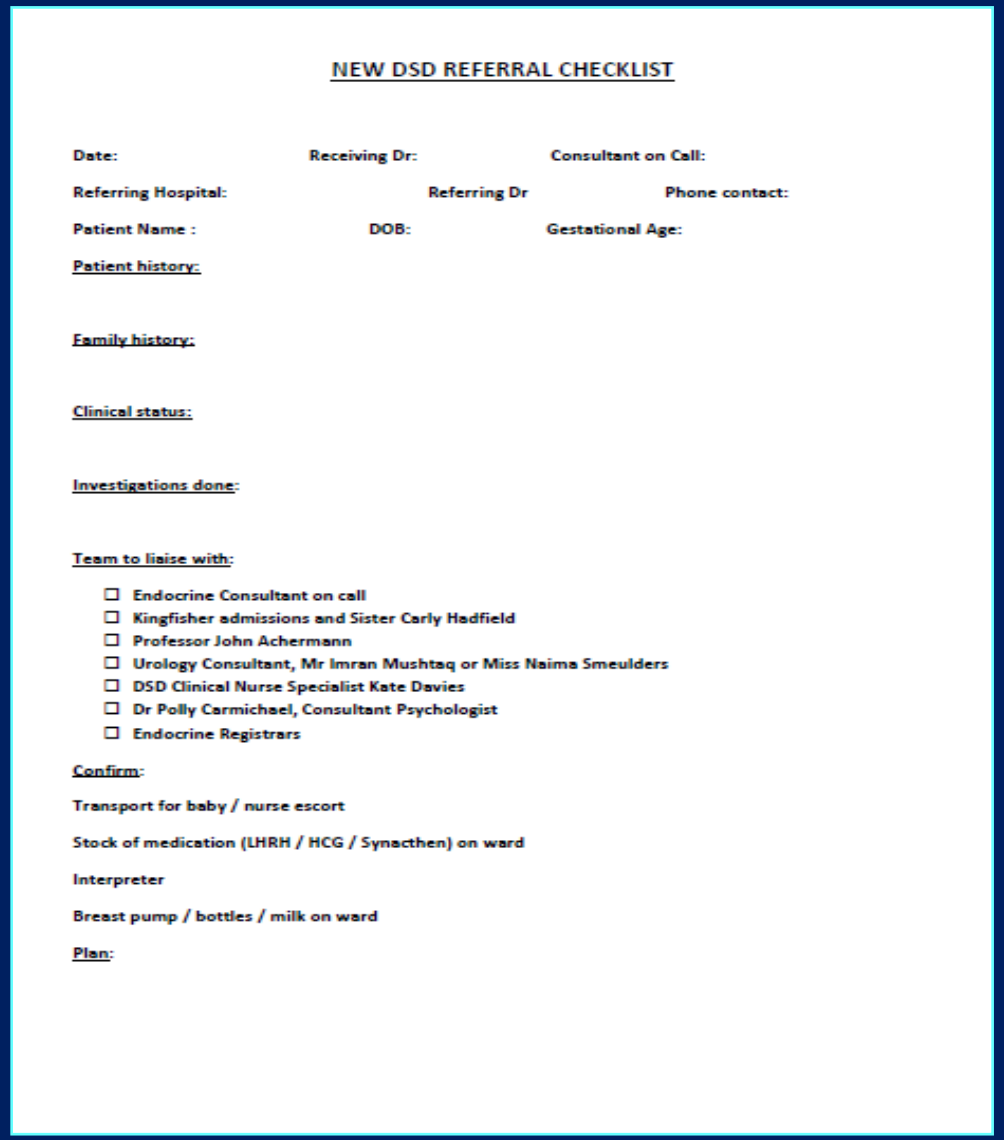
- CHARGE syndrome
- Testicular tumour
- Duplicated genitalia
- Aphalia
- Delayed / precocious puberty
- SOX2, SOX9
- Smith – Lemli – Opitz
- Meacham syndrome
- Lipoid CAH

How we get a referral

- On call registrar receives a telephone call from referring Doctor from another hospital
- Alerts main DSD team
- Plan admission
  - Within 5 days if newborn
- Asks referring Dr to undertake specific investigations
- Plans investigations
  - Pelvic ultrasound, medical photography
- Clinical Nurse Specialist..

DSD referral form

- Emailed to lead consultant
- Filed in patient notes
- Filed in DSD file



NEW DSD REFERRAL CHECKLIST

Date: **Receiving Dr:** **Consultant on Call:**

Referring Hospital: **Referring Dr** **Phone contact:**

Patient Name : **DOB:** **Gestational Age:**

Patient history:

Family history:

Clinical status:

Investigations done:

Team to liaise with:

- ☐ Endocrine Consultant on call
- ☐ Kingfisher admissions and Sister Carly Hadfield
- ☐ Professor John Achermann
- ☐ Urology Consultant, Mr Imran Mushtaq or Miss Naima Smeulders
- ☐ DSD Clinical Nurse Specialist Kate Davies
- ☐ Dr Polly Carmichael, Consultant Psychologist
- ☐ Endocrine Registrars

Confirm:

Transport for baby / nurse escort

Stock of medication (LHRH / HCG / Synacthen) on ward

Interpreter

Breast pump / bottles / milk on ward

Plan:

CNS role for new admission

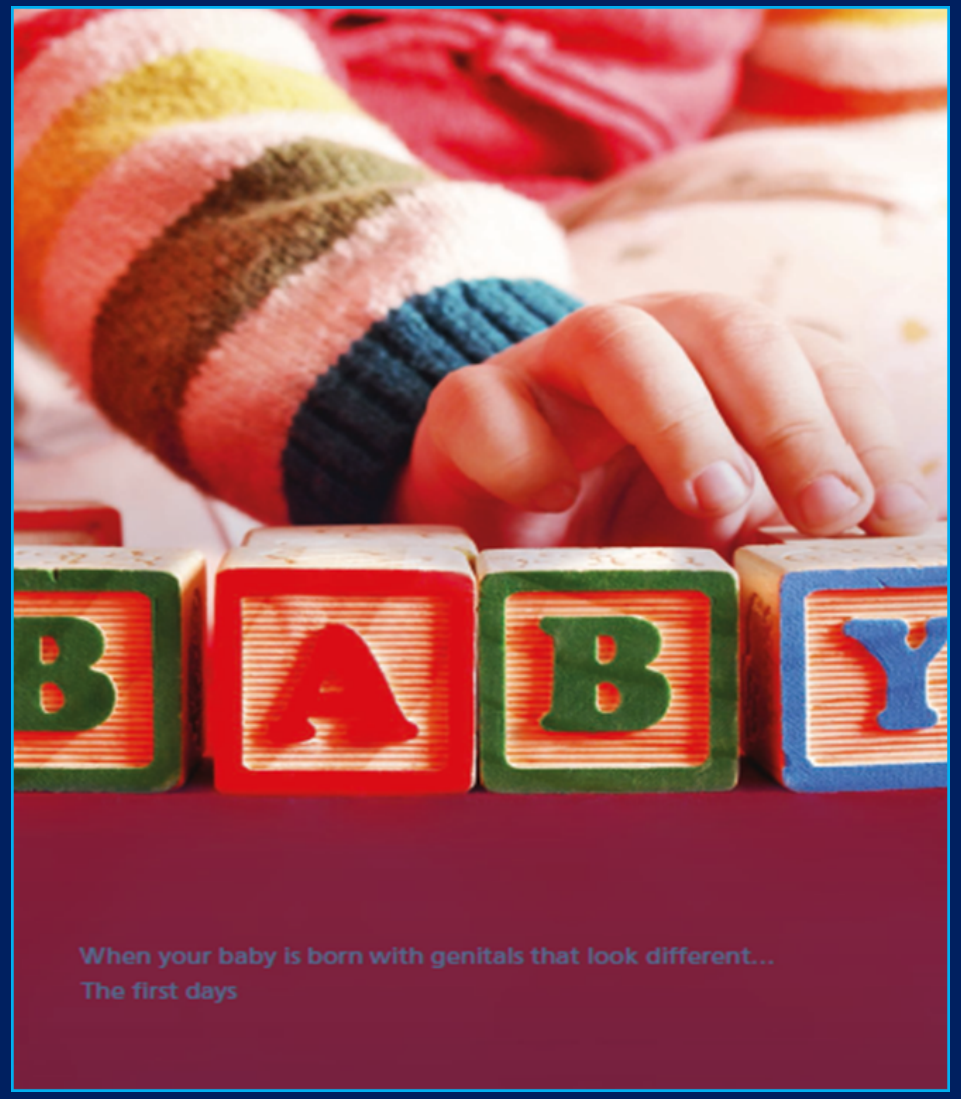
- Ensure referring team has parents admission leaflet
- Ensure Registrar has completed referral form
- Liaise with:
  - Consultant Endocrinologist
  - Psychologist
  - Consultant Urologist
  - Sister and admissions team on Kingfisher ward
  - PARENTS!
    - Admissions leaflet from GOSH website (under review)
- Set a date and time for one day admission

It’s the little things..

- How is the baby feeding
  - Breast pump, bottles, quiet area available
  - Bottle feeding – enough milk
- Ensure parents bring
  - Phone chargers, nappies, wipes, books etc, lists of questions
  - Maternity notes, child health care notes, referral letters
  - Money for parking
- Can they speak English
  - Arrange interpreter, prepare translated information

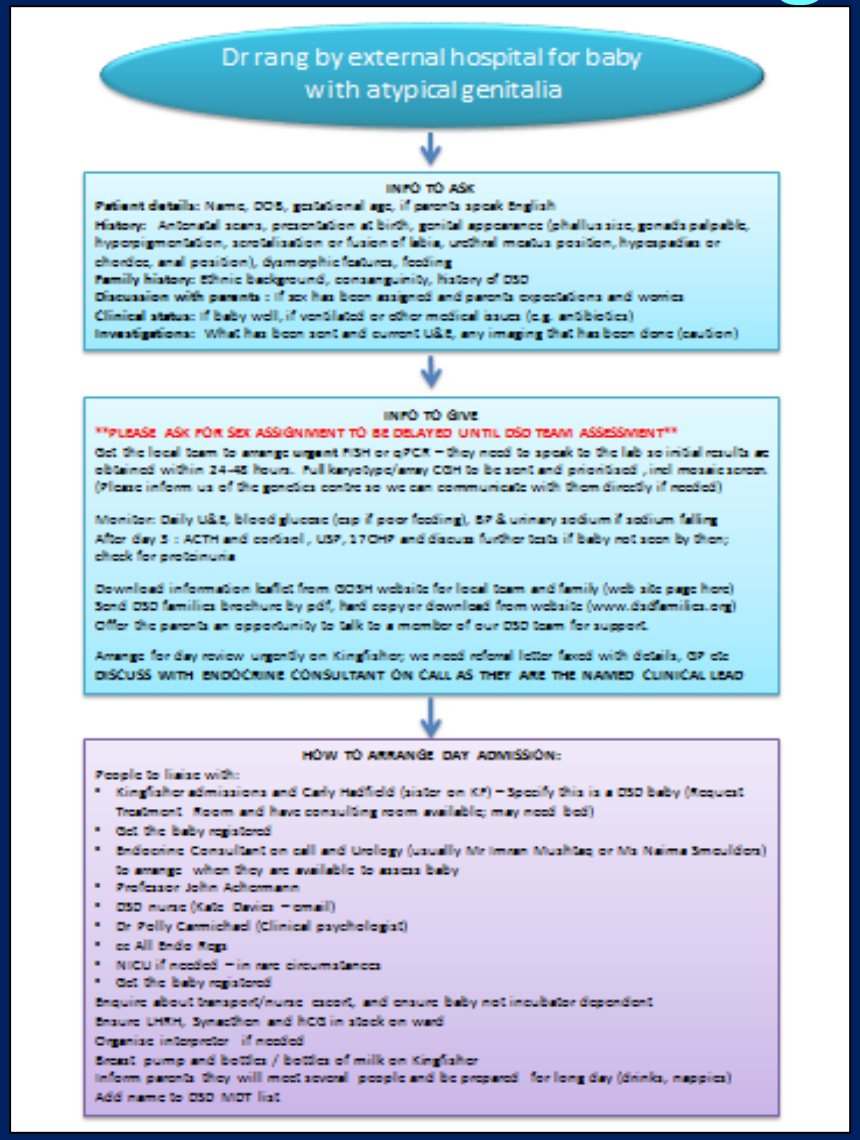
- Transport / nurse escort
  - Liaise with NICU / Respiratory ward (Badger)
- Ensure GnRH, Synacthen and HCG in stock on ward if need be
- Prepare information packs for parents
  - DSD families leaflet
  - Cortisol deficiency booklets
  - CAH information
  - CNS business card / contact details

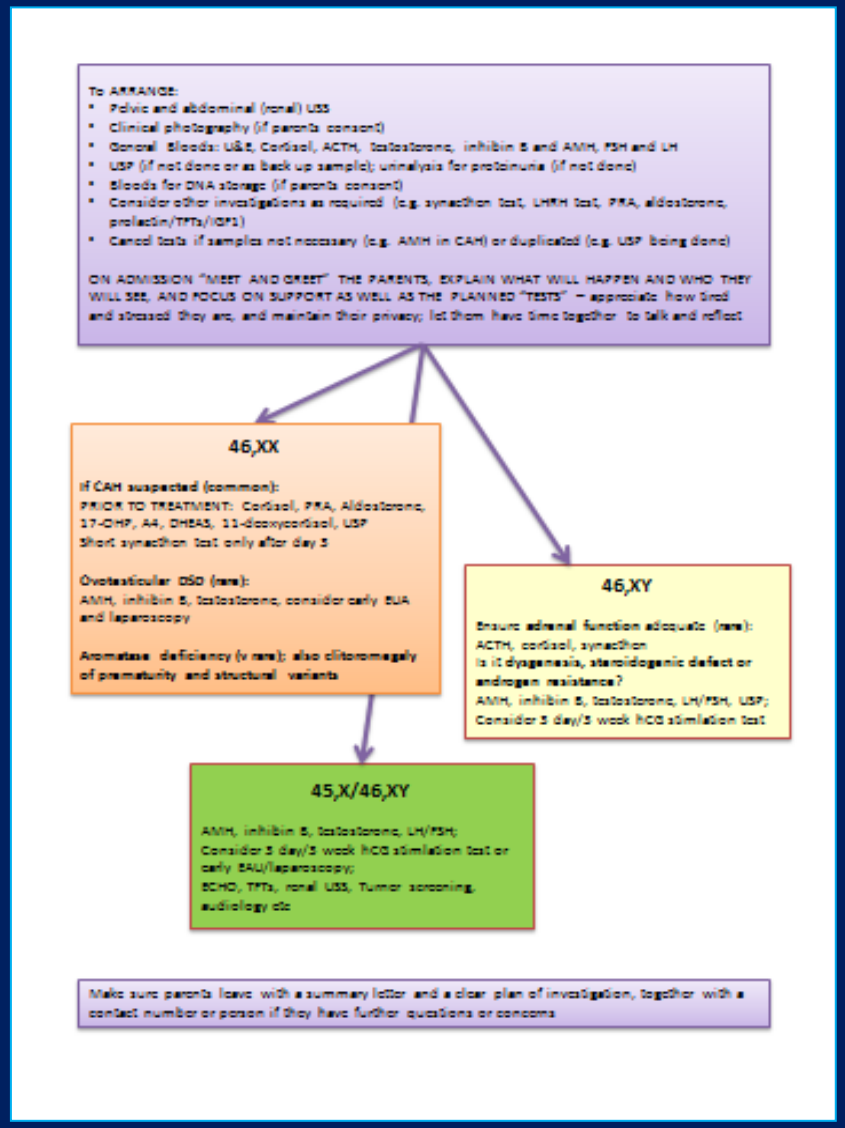
DSD families leaflet



- dsdfamilies.org
- UK based support group
  - Information and support resource for families with children, teens and young adults with a DSD
  - Links to other support groups throughout the UK
    - CAH, TS, Hypospadias, Klinefelter, AIS
  - Links to international DSD support groups

GOSH Diagnostic flow chart





Flow chart broken down…

Dr rang by external hospital for baby with atypical genitalia

INFO TO ASK

Patient details: Name, DOB, gestational age, if parents speak English

History: Antenatal scans, presentation at birth, genital appearance (phallus size, gonads palpable, hyperpigmentation, scrotalisation or fusion of labia, urethral meatus position, hypospadias or chordee, anal position), dysmorphic features, feeding

Family history: Ethnic background, consanguinity, history of DSD

Discussion with parents: If sex has been assigned and parents expectations and worries

Clinical status: If baby well, if ventilated or other medical issues (e.g. antibiotics)

Investigations: What has been sent and current U&E, any imaging that has been done (caution)

INFO TO GIVE

****PLEASE ASK FOR SEX ASSIGNMENT TO BE DELAYED UNTIL DSD TEAM ASSESSMENT****

Get the local team to arrange **urgent** FISH or qPCR – they need to speak to the lab so initial results are obtained within 24-48 hours. Full karyotype/array CGH to be sent and prioritised , incl mosaic screen. (Please inform us of the genetics centre so we can communicate with them directly if needed)

Monitor: Daily U&E, blood glucose (esp if poor feeding), BP & urinary sodium if sodium falling
After day 3 : ACTH and cortisol , USP, 17OHP and discuss further tests if baby not seen by then; check for proteinuria

Download information leaflet from GOSH website for local team and family (web site page here)
Send DSD families brochure by pdf, hard copy or download from website (www.dsdfamilies.org)
Offer the parents an opportunity to talk to a member of our DSD team for support.

Arrange for day review urgently on Kingfisher; we need referral letter faxed with details, GP etc
DISCUSS WITH ENDOCRINE CONSULTANT ON CALL AS THEY ARE THE NAMED CLINICAL LEAD

HOW TO ARRANGE DAY ADMISSION:

People to liaise with:

- Kingfisher admissions and Carly Hadfield (sister on KF) – Specify this is a DSD baby (Request Treatment Room and have consulting room available; may need bed)
- Get the baby registered
- Endocrine Consultant on call and Urology (usually Mr Imran Mushtaq or Ms Naima Smeulders) to arrange when they are available to assess baby
- Professor John Achermann
- DSD nurse (Kate Davies – email)
- Dr Polly Carmichael (Clinical psychologist)
- cc All Endo Regs
- NICU if needed – in rare circumstances
- Get the baby registered

Enquire about transport/nurse escort, and ensure baby not incubator dependent

Ensure LHRH, Synacthen and hCG in stock on ward

Organise interpreter if needed

Breast pump and bottles / bottles of milk on Kingfisher

Inform parents they will meet several people and be prepared for long day (drinks, nappies)

Add name to DSD MDT list

To ARRANGE:

- Pelvic and abdominal (renal) USS
- Clinical photography (if parents consent)
- General Bloods: U&E, Cortisol, ACTH, testosterone, inhibin B and AMH, FSH and LH
- USP (if not done or as back up sample); urinalysis for proteinuria (if not done)
- Bloods for DNA storage (if parents consent)
- Consider other investigations as required (e.g. synacthen test, LHRH test, PRA, aldosterone, prolactin/TFTs/IGF1)
- Cancel tests if samples not necessary (e.g. AMH in CAH) or duplicated (e.g. USP being done)

ON ADMISSION "MEET AND GREET" THE PARENTS, EXPLAIN WHAT WILL HAPPEN AND WHO THEY WILL SEE, AND FOCUS ON SUPPORT AS WELL AS THE PLANNED "TESTS" – appreciate how tired and stressed they are, and maintain their privacy; let them have time together to talk and reflect

46,XX

If CAH suspected (common):
PRIOR TO TREATMENT: Cortisol, PRA, Aldosterone, 17-OHP, A4, DHEAS, 11-deoxycortisol, USP
Short synacthen test only after day 3

Ovotesticular DSD (rare):
AMH, inhibin B, testosterone, consider early EUA and laparoscopy

Aromatase deficiency (v rare); also clitoromegaly of prematurity and structural variants

45,X/46,XY

AMH, inhibin B, testosterone, LH/FSH;
Consider 3 day/3 week hCG stimlation test or early EAU/laparoscopy;
ECHO, TFTs, renal USS, Turner screening, audiology etc

46,XY

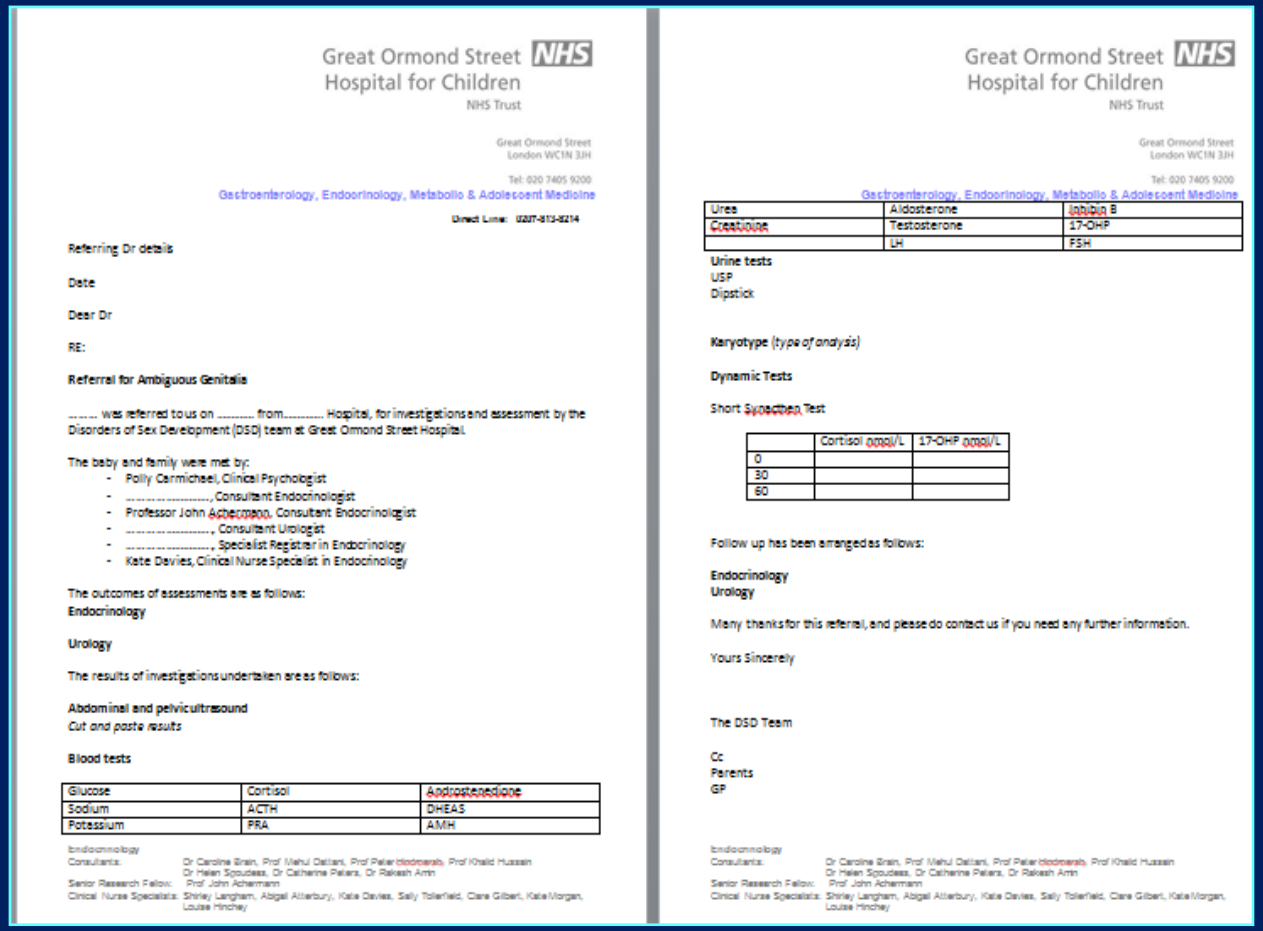
Ensure **adrenal function** adequate (**rare**):
ACTH, cortisol, synacthen
Is it **dysgenesis, steroidogenic defect or androgen resistance**?
AMH, inhibin B, testosterone, LH/FSH, USP;
Consider 3 day/3 week hCG stimlation test

On the day

- Parents and child come to Kingfisher ward
- CNS greets family
  - Shows them their room
  - Ensures comfort, privacy
  - Explains procedures
- MDT appointments may be spread out
- CNS
  - Answer questions
  - Works with Registrar
  - Clinical examination
  - Weight, length, observations etc

On discharge..

- Template discharge letter
- Given to family at end of day
  - Details MDT meeting
  - Decision re sex of rearing
  - Investigations carried out
  - Differential diagnoses
- If CAH
  - Appt for adrenal NLC 1/12 later
  - Liaise with local hospital, nursery, community nurses , ambulance services..



Great Ormond Street NHS
Hospital for Children
NHS Trust

Great Ormond Street
London WC1N 3JH

Tel: 020 7405 9200

Gastroenterology, Endocrinology, Metabolic & Adolescent Medicine

Direct Line: 0207-813-8214

Referring Dr details

Date

Dear Dr

RE:

Referral for Ambiguous Genitalia

..... was referred to us on ..... from..... Hospital, for investigations and assessment by the Disorders of Sex Development (DSD) team at Great Ormond Street Hospital.

The baby and family were met by:

- Polly Carmichael, Clinical Psychologist
- ....., Consultant Endocrinologist
- Professor John Achermann, Consultant Endocrinologist
- ....., Consultant Urologist
- ....., Specialist Registrar in Endocrinology
- Kate Davies, Clinical Nurse Specialist in Endocrinology

The outcomes of assessments are as follows:

Endocrinology

Urology

The results of investigations undertaken are as follows:

Abdominal and pelvic ultrasound

Cut and paste results

Blood tests

Glucose	Cortisol	Androstenedione
Sodium	ACTH	DHEAS
Potassium	PRA	AMH

Endocrinology
Consultants: Dr Caroline Brain, Prof Mehul Dattani, Prof Peter Clayton, Prof Khalid Hussain, Dr Helen Spoudeas, Dr Catherine Peters, Dr Rakesh Amin
Senior Research Fellow: Prof John Achermann
Clinical Nurse Specialists: Shirley Langham, Abigail Atterbury, Kate Davies, Sally Tollerfield, Clare Gilbert, Kate Morgan, Louise Hinchey

Great Ormond Street NHS
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Great Ormond Street
London WC1N 3JH

Tel: 020 7405 9200

Gastroenterology, Endocrinology, Metabolic & Adolescent Medicine

Urea	Aldosterone	Inhibin B
Creatinine	Testosterone	17-OHP
	LH	FSH

Urine tests
USP
Dipstick

Karyotype (*type of analysis*)

Dynamic Tests

Short Synacthen Test

	Cortisol (nmol/L)	17-OHP (nmol/L)
0		
30		
60		

Follow up has been arranged as follows:

Endocrinology
Urology

Many thanks for this referral, and please do contact us if you need any further information.

Yours Sincerely

The DSD Team

Cc
Parents
GP

Endocrinology
Consultants: Dr Caroline Brain, Prof Mehul Dattani, Prof Peter Clayton, Prof Khalid Hussain, Dr Helen Spoudeas, Dr Catherine Peters, Dr Rakesh Amin
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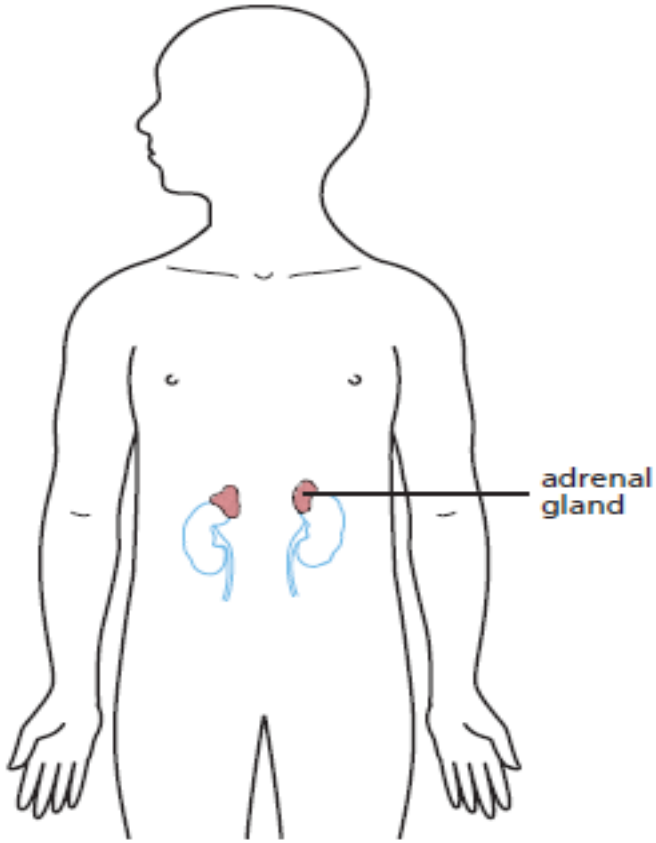
Congenital adrenal hyperplasia (CAH)

This information sheet from Great Ormond Street Hospital (GOSH) explains about the medical condition congenital adrenal hyperplasia (CAH) and what to expect when your child comes to GOSH for assessment and treatment.



Congenital adrenal hyperplasia is group of inherited conditions that are present at birth (congenital) where the adrenal gland is larger than usual (hyperplasia). In CAH, the body is missing an enzyme (chemical substance) that stimulates the adrenal glands to release the cortisol hormone. Lacking this hormone means that the body is less able to cope with stress, either emotionally or physically, which can be life threatening. It also makes the level of androgen (male hormone) increase, which causes male characteristics to appear early in boys or inappropriately in girls.

The adrenal glands rest on the tops of the kidneys. They are part of the endocrine system, which organises the release of hormones within the body. Hormones are chemical messengers that switch on and off processes within the body.

The adrenal glands consist of two parts:

- the medulla (inner section) which makes the hormone 'adrenaline' which is part of the 'fight or flight' response a person has when stressed. This is not usually affected in CAH.

GP letter on day of discharge

GP details

Date

Dear Dr

RE:

Diagnosis: Congenital Adrenal Hyperplasia

…… was referred to us on ……. from……. Hospital, and a diagnosis of Congenital Adrenal Hyperplasia has been made, and s/he has cortisol deficiency.

She/he has been commenced on the following medication and we would be very grateful if you could commence a repeat prescription for:

Hydrocortisone …… mg (as Hydrocortisone 10mg tablets, **NOT** suspension)
Fludrocortisone …….. mcg daily
Oral salt supplements (until one year of age) in the 5mmol/ml 30% Sodium Chloride solution – 5mmols/kg/day, in 4 divided doses = ……mls per dose four times a day

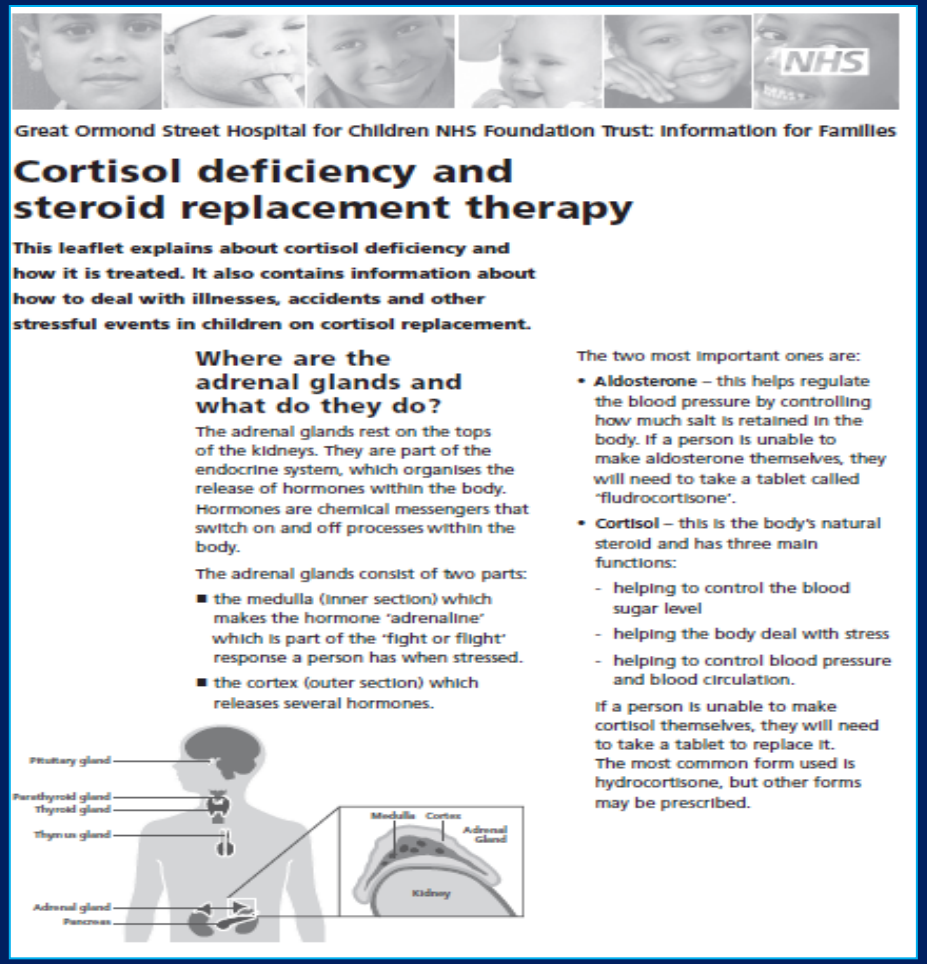
Also to be included on his/her prescription:
Hydrocortisone Emergency Pack to be renewed yearly:
Efcortesol 100mg vials, 25/50/100mg to be given IM in an emergency.
Glucose Gel 25g tube, 1/3 tube orally to be given in an emergency.

We have given ……'s parents a tablet cutter and a tablet crusher, and have educated them in how to prepare and administer their medication. ……'s parents have had education in his/her management during times of illness and they have been trained in giving IM hydrocortisone, 25/50/100mg, should the need arise, along with oral glucose gel. A steroid card has been given, and also information about Medic Alert jewellery to start wearing as soon as possible.

We have arranged with the local hospital and the local Paediatrician (……) to have fast track access should he/she require emergency IM hydrocortisone. We have also set up an arrangement with ……Ambulance Service to ensure a red alert system is in place.

- Medication onto repeat prescription system
- Tablets not suspension
- Also highlights that the baby can have all of their usual childhood immunisations

Sick day and emergency management



Great Ormond Street Hospital for Children NHS Foundation Trust: Information for Families

Cortisol deficiency and steroid replacement therapy

This leaflet explains about cortisol deficiency and how it is treated. It also contains information about how to deal with illnesses, accidents and other stressful events in children on cortisol replacement.

Where are the adrenal glands and what do they do?

The adrenal glands rest on the tops of the kidneys. They are part of the endocrine system, which organises the release of hormones within the body. Hormones are chemical messengers that switch on and off processes within the body.

The adrenal glands consist of two parts:

- the medulla (inner section) which makes the hormone 'adrenaline' which is part of the 'fight or flight' response a person has when stressed.
- the cortex (outer section) which releases several hormones.

The two most important ones are:

- **Aldosterone** – this helps regulate the blood pressure by controlling how much salt is retained in the body. If a person is unable to make aldosterone themselves, they will need to take a tablet called 'fludrocortisone'.
- **Cortisol** – this is the body's natural steroid and has three main functions:
  - helping to control the blood sugar level
  - helping the body deal with stress
  - helping to control blood pressure and blood circulation.

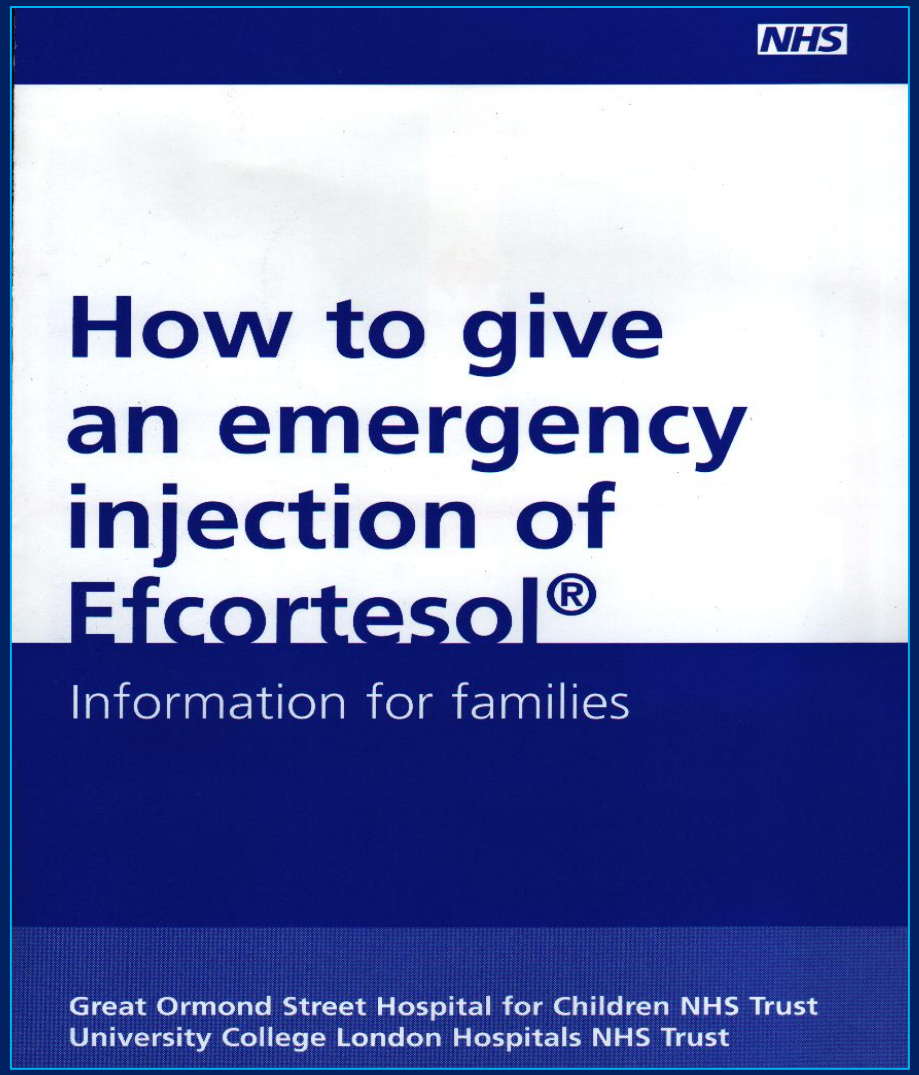
If a person is unable to make cortisol themselves, they will need to take a tablet to replace it. The most common form used is hydrocortisone, but other forms may be prescribed.

- Doubling up on hydrocortisone when unwell
- Additional 4am dose (same as morning dose)

Sick day and emergency management

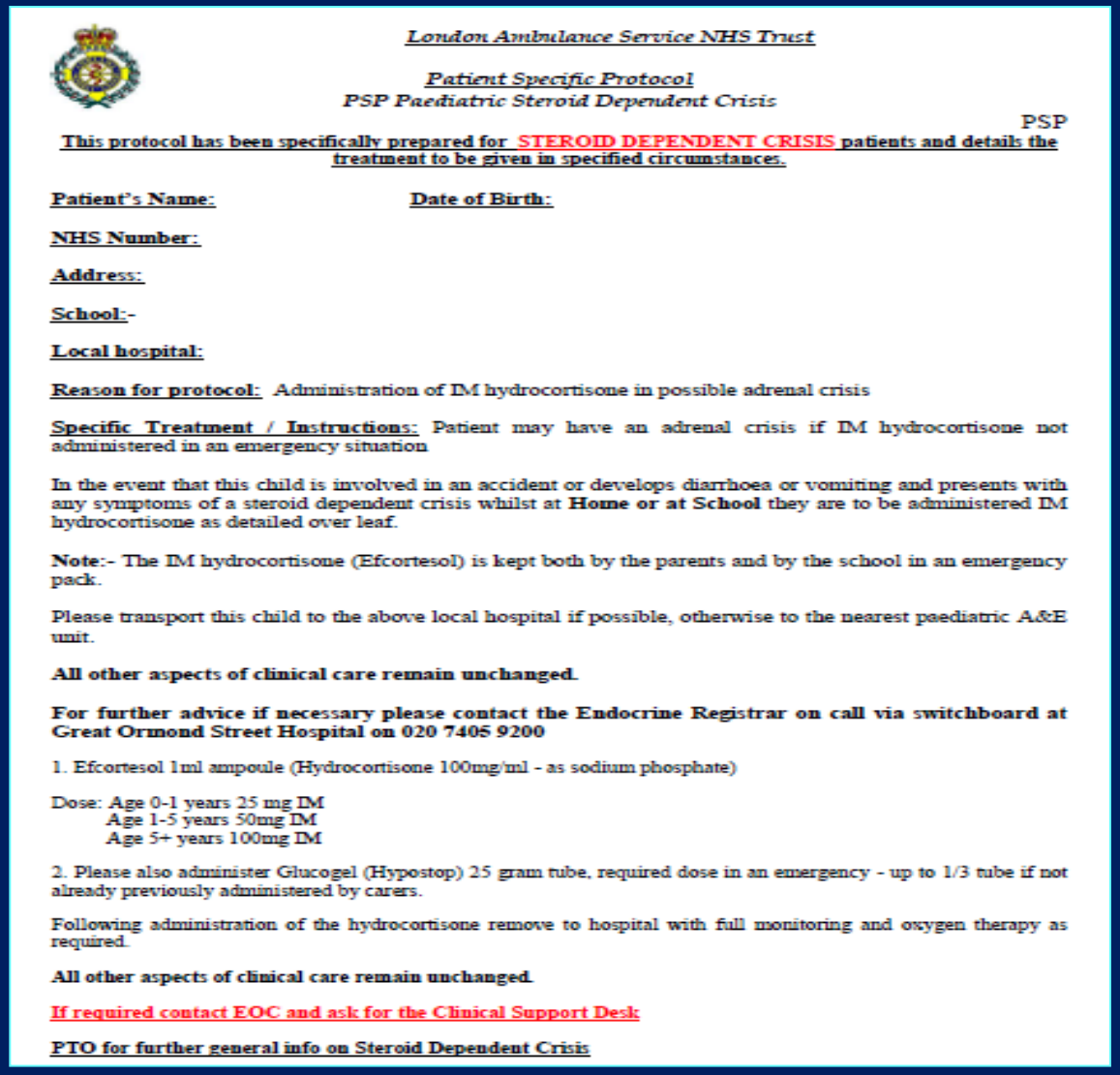
- Emergency injection of hydrocortisone and oral glucogel
- Liaise with nurseries
  - Schools when older
- Medic alert jewellery

- Usually dispense x2 emergency packs
  - Home
  - Bag
    - Another when older for nursery / school



Emergency services

- Contact details for all UK ambulance services
- Red flag system



London Ambulance Service NHS Trust

Patient Specific Protocol
PSP Paediatric Steroid Dependent Crisis

PSP

This protocol has been specifically prepared for STEROID DEPENDENT CRISIS patients and details the treatment to be given in specified circumstances.

Patient's Name: **Date of Birth:**

NHS Number:

Address:

School:-

Local hospital:

Reason for protocol: Administration of IM hydrocortisone in possible adrenal crisis

Specific Treatment / Instructions: Patient may have an adrenal crisis if IM hydrocortisone not administered in an emergency situation

In the event that this child is involved in an accident or develops diarrhoea or vomiting and presents with any symptoms of a steroid dependent crisis whilst at **Home or at School** they are to be administered IM hydrocortisone as detailed over leaf.

Note:- The IM hydrocortisone (Efcortesol) is kept both by the parents and by the school in an emergency pack.

Please transport this child to the above local hospital if possible, otherwise to the nearest paediatric A&E unit.

All other aspects of clinical care remain unchanged.

For further advice if necessary please contact the Endocrine Registrar on call via switchboard at Great Ormond Street Hospital on 020 7405 9200

1. Efcortesol 1ml ampoule (Hydrocortisone 100mg/ml - as sodium phosphate)

Dose: Age 0-1 years 25 mg IM
Age 1-5 years 50mg IM
Age 5+ years 100mg IM

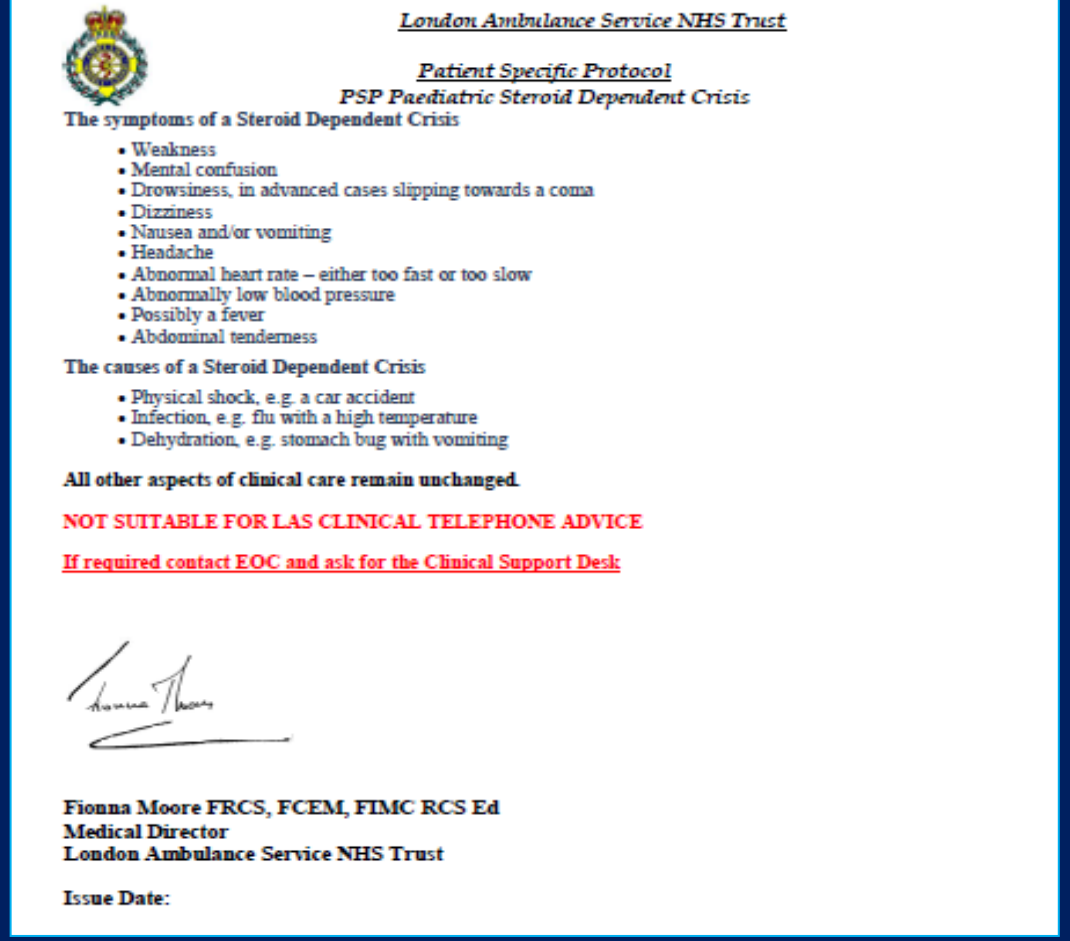
2. Please also administer Glucogel (Hypostop) 25 gram tube, required dose in an emergency - up to 1/3 tube if not already previously administered by carers.

Following administration of the hydrocortisone remove to hospital with full monitoring and oxygen therapy as required.

All other aspects of clinical care remain unchanged.

If required contact EOC and ask for the Clinical Support Desk

PTO for further general info on Steroid Dependent Crisis



London Ambulance Service NHS Trust

Patient Specific Protocol
PSP Paediatric Steroid Dependent Crisis

The symptoms of a Steroid Dependent Crisis

- Weakness
- Mental confusion
- Drowsiness, in advanced cases slipping towards a coma
- Dizziness
- Nausea and/or vomiting
- Headache
- Abnormal heart rate – either too fast or too slow
- Abnormally low blood pressure
- Possibly a fever
- Abdominal tenderness

The causes of a Steroid Dependent Crisis

- Physical shock, e.g. a car accident
- Infection, e.g. flu with a high temperature
- Dehydration, e.g. stomach bug with vomiting

All other aspects of clinical care remain unchanged.

NOT SUITABLE FOR LAS CLINICAL TELEPHONE ADVICE

If required contact EOC and ask for the Clinical Support Desk

Fionna Moore FRCS, FCEM, FIMC RCS Ed
Medical Director
London Ambulance Service NHS Trust

Issue Date:

Emergency hospital letter

Great Ormond Street Hospital for Children NHS Trust

Great Ormond Street
London WC1N 3JH

Tel: 020 7405 9200
Gastroenterology, Endocrinology, Metabolic & Adolescent Medicine (GEMA)
Direct Line: 0207-813-8214

Re:
Diagnosis:
Medications: Hydrocortisone (oral)
Fludrocortisone (oral)
NaCl supplements 5mmol/ml 30% solution:

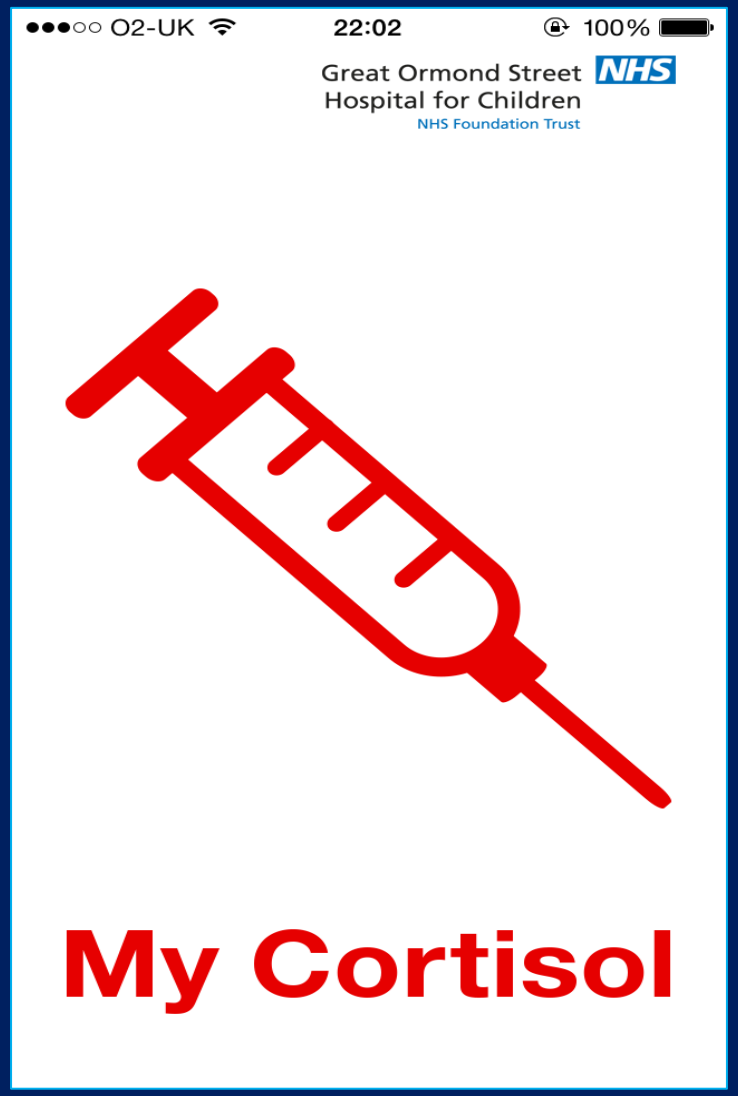
Instructions for Hospital Doctor

In view of this patients cortisol deficiency, if this patient is brought to hospital as an emergency, the following management is advised:

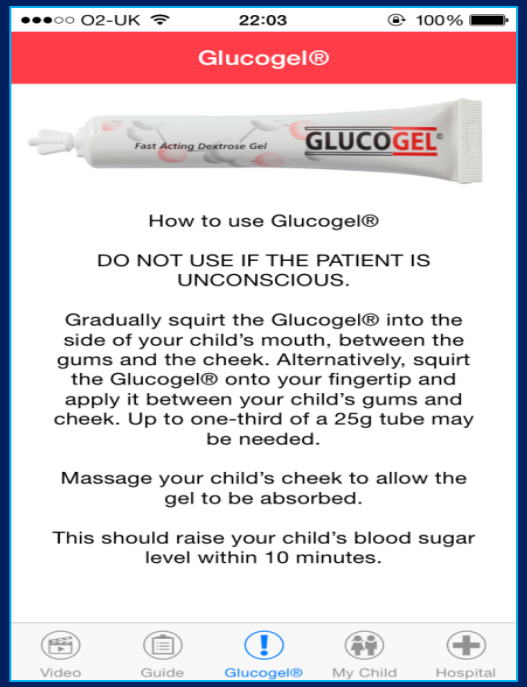
- If patient drowsy and unresponsive give IM hydrocortisone in the following doses immediately (0-1yr – 25mgs; 1-5 yrs – 50mgs; > 5yrs – 100mgs) if patient has not already had IM hydrocortisone administered by ambulance crew or parents.
- Take blood for U&Es, glucose and osmolality
- If blood glucose is < 2.5 mmol, give bolus of 2mg/kg 10% dextrose
- If patient is drowsy, hypotensive and peripherally shut down, give 20ml/kg of normal saline, insert an IV cannula and then continue with usual dextrose saline infusion
- Continue with bolus IV hydrocortisone at 2mg/kg every 4 hours until patient is tolerating oral fluids and then swap to double usual oral Hydrocortisone doses until patient fully recovered and back to normal self (usually 2-3 days on double usual hydrocortisone doses).
- **Important:** Please admit for a minimum of 12 hours

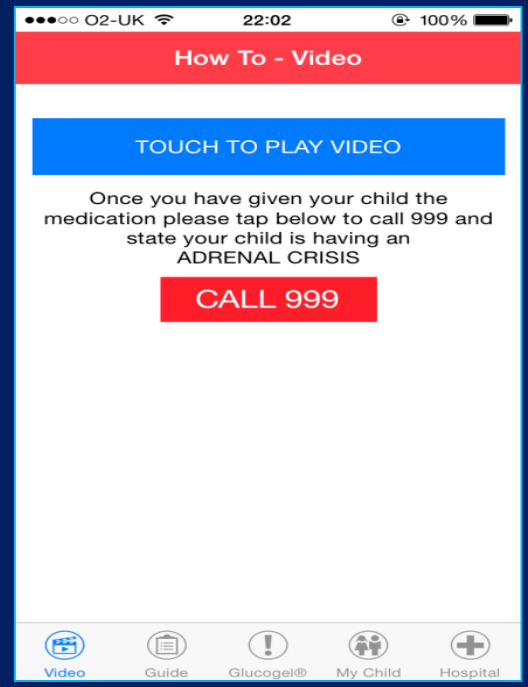
If there is any doubt about this patients management, advice can be obtained via Great Ormond Street Hospital switchboard (0207 405 9200, asking for the Endocrine Registrar on Call).

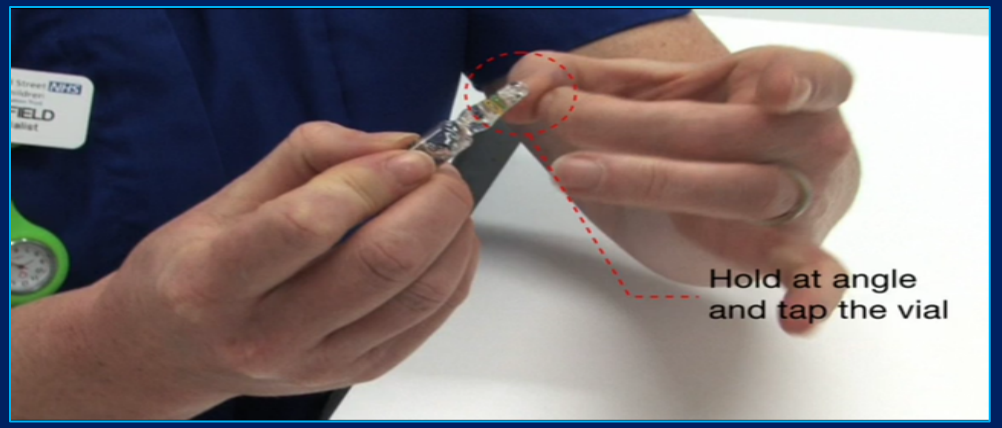
My Cortisol App

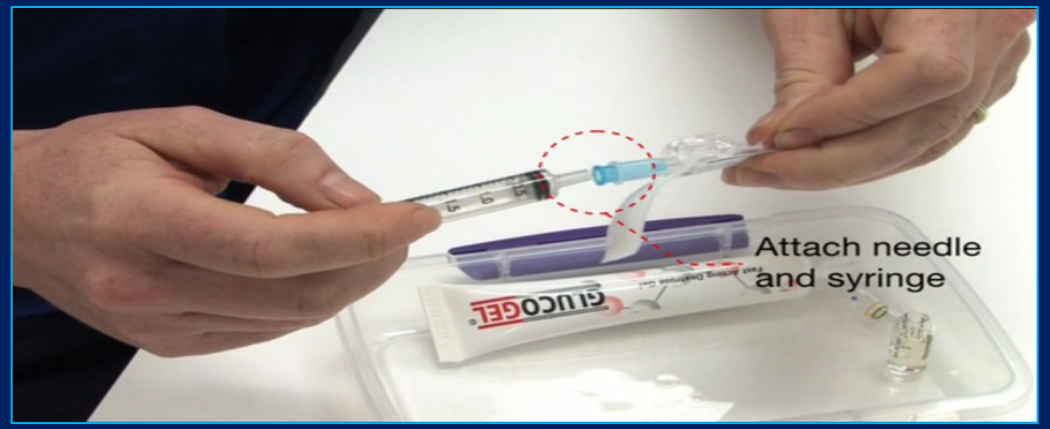



O2-UK 22:02 100%
How To - Guide
How to give an emergency Efcortesol® injection
1
Syringe
Needle
Efcortesol
Equipment
2
Tap
Break
Open the vial
3
Attach needle & syringe
4
Draw up
5
6
0.25ml
0.5ml
1ml
Video
Guide
Glucogel®
My Child
Hospital


O2-UK 22:03 100%
Glucogel®
Fast Acting Dextrose Gel
GLUCOGEL
How to use Glucogel®
DO NOT USE IF THE PATIENT IS UNCONSCIOUS.
Gradually squirt the Glucogel® into the side of your child's mouth, between the gums and the cheek. Alternatively, squirt the Glucogel® onto your fingertip and apply it between your child's gums and cheek. Up to one-third of a 25g tube may be needed.
Massage your child's cheek to allow the gel to be absorbed.
This should raise your child's blood sugar level within 10 minutes.
Video
Guide
Glucogel®
My Child
Hospital


O2-UK 22:02 100%
How To - Video
TOUCH TO PLAY VIDEO
Once you have given your child the medication please tap below to call 999 and state your child is having an ADRENAL CRISIS
CALL 999
Video
Guide
Glucogel®
My Child
Hospital


Hold at angle
and tap the vial


Attach needle
and syringe
GLUCOGEL

Steroid card

Instructions for Hospital Doctor

Dear Doctor,

If this patient is brought to hospital as an emergency the following management is advised:

1) Insert an IV cannula
2) Take blood for U&Es, glucose, and perform any other appropriate tests (e.g. urine culture)
3) Check capillary blood glucose level
4) Give 100 mg hydrocortisone intravenously as bolus (unnecessary if patient has already been given IM hydrocortisone)
5) Commence IV infusion of 0.45% sodium chloride and 5% glucose at maintenance rate (extra if patient is dehydrated). Add potassium depending on electrolyte
6) Commence hydrocortisone infusion (50 mg hydrocortisone in 50ml 0.9% sodium chloride via syringe pump)
7) Monitor for at least twelve hours before discharge

IMPORTANT! If blood glucose is < 2.5 mmol/l, give bolus of 2 ml/kg of 10% glucose

If patient is drowsy, hypotensive and peripherally shut down with poor capillary return give 20ml/kg of 0.9% sodium chloride stat.

If in any doubt about this patient's management, please contact the urgent advice numbers

Useful Contact Numbers:

GOSH Switchboard
Tel: 020 7405 9200

For Urgent Advice:
Tel: 020 7405 9200 and ask to be put through to the endocrine registrar on call

University College Hospital Switchboard
Tel: 0845 155 5000

For Urgent Advice:
Tel: 0845 155 5000 and ask to be put through to the endocrine registrar on call.

Great Ormond Street Hospital for Children NHS Trust and University College London Hospitals NHS Foundation Trust NHS

CORTISOL DEFICENCY

THE OWNER OF THIS CARD IS ON CORTISOL REPLACEMENT THERAPY

Name _____

_____

Address _____

_____

_____

_____

Tel _____

Mobile: _____

Date of Birth ____/____/____

Hospital No _____

Affix photo here

Consultant _____

Hospital _____

Address _____ _____

Tel _____ Fax _____

General Practitioner _____

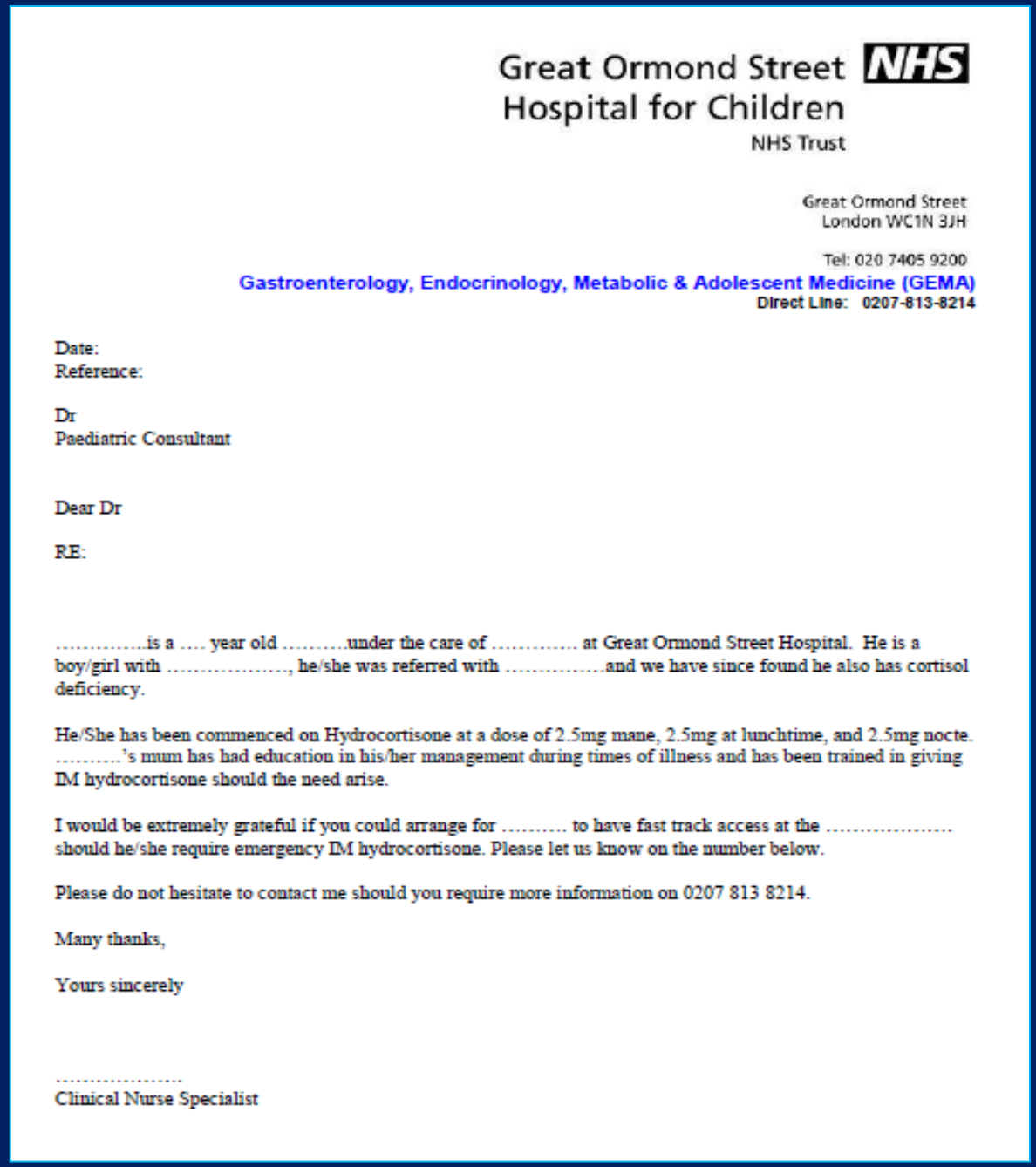
Address _____

Tel _____ Fax _____

Great Ormond Street Hospital for Children NHS Trust and University College London Hospitals NHS Foundation Trust NHS

Primary Care

- Open access onto local paediatric ward
- Contact details for all UK childrens' community nursing teams
- Medical team to liaise with local medical team to arrange formal shared care plan



Great Ormond Street Hospital for Children NHS Trust

Great Ormond Street
London WC1N 3JH

Tel: 020 7405 9200
Gastroenterology, Endocrinology, Metabolic & Adolescent Medicine (GEMA)
Direct Line: 0207-813-8214

Date:
Reference:

Dr
Paediatric Consultant

Dear Dr

RE:

……..is a …. year old …….under the care of …… at Great Ormond Street Hospital. He is a boy/girl with ……., he/she was referred with ……..and we have since found he also has cortisol deficiency.

He/She has been commenced on Hydrocortisone at a dose of 2.5mg mane, 2.5mg at lunchtime, and 2.5mg nocte. ……'s mum has had education in his/her management during times of illness and has been trained in giving IM hydrocortisone should the need arise.

I would be extremely grateful if you could arrange for ……. to have fast track access at the …… should he/she require emergency IM hydrocortisone. Please let us know on the number below.

Please do not hesitate to contact me should you require more information on 0207 813 8214.

Many thanks,

Yours sincerely

……
Clinical Nurse Specialist

Primary care – blood levels

Cortisol bloods plan for new CAH baby

Discharge following birth

Week 1

Week 2

Week 3

Week 4

Week 6

Week 8

Week 10

Week 12 / Month 3

Month 4

Month 5

Month 6

Month 7

Month 8

Month 9

Month 10

Month 11

Month 12

On discharge following birth: weekly bloods for 4 weeks.

2 weekly for the next 8 weeks.

4 weekly until fully weaned.

When weaned (around age 1yr) - check 4 weeks later, and then at annual reviews in clinic / 6 monthly.

- Liaise with local teams for community nurses to visit family and take regular bloods for U&E
- Ensure results are fed back to GOSH

Adrenal nurse led clinic

- First appointment
  - One month after diagnosis / discharge from GOSH
  - Discuss
    - Compliance
    - Management of medication
    - Re-educate sick day and emergency management
      - Teach injection technique
  - Follow up on any queries the family have
  - Liaise with Urology if female
  - Liaise with local teams for recent blood results
  - Discuss patient support groups

CNS Roles

- Hamric & Spross (1989):
- **Consultant**
  - CNS as a resource or a consultant
- **Educator**
  - Educating staff in disease specifics
  - Educating patients in self-care management
- **Researcher**
  - Involvement in clinical trials
  - CNS's own research related to nursing policy and practice
- **Collaborator**
  - Importance of MD teamwork
- **Leader**
  - Leadership and management

- **Change agent** *(Miller, 1995)*
- **Advocate** *(Miller, 1995)*
- **Liaison** *(Gibson and Bamford, 2001)*
- **Communicator-Carer** *(McCreaddie, 2001)*
- **Entrepreneurs** *(Austin, 2006)*
  - Visualising how clinical services should function

Clinical Nurse Specialist Roles

- **Clinical Expert**
  - How can your knowledge and skills within endocrinology be enhanced
    - Further training / development
- **Education**
  - Sub role as Educator
    - Patients, families, staff
  - Evaluate and develop educational programmes
  - Build teaching packages for patient education
    - Eg IM HC, GH devices, puberty

- **Consultant**
  - Leading on case management
  - Becoming more involved in external forums
    - Advisory boards, society committees
- **Patient advocate**
  - Identify patient support groups not already utilised by your team
    - Develop and strengthen links
    - Develop own patient literature

Clinical Nurse Specialist Roles

- **Research**
  - Identify gaps in your service which could use research / audit to prove shortfalls
    - Or even *positive* aspects
  - Patient questionnaires
    - Satisfaction in patient pathways
- **Collaborator**
  - Enhance collaboration within the MDT and interdisciplinary
    - Ensure common purpose
  - Build and develop relationships with outside personnel
- **Leadership / Management**
  - Lead in developing and attaining team goals
    - Contribute to practice development
    - Develop patient care pathways
      - Patient literature
- **Change Agent**
  - Provide evidence where CNS intervention could be useful
    - Nurse led clinics
    - Telephone clinics
  - Suggest, develop and implement business plans

Specifics..

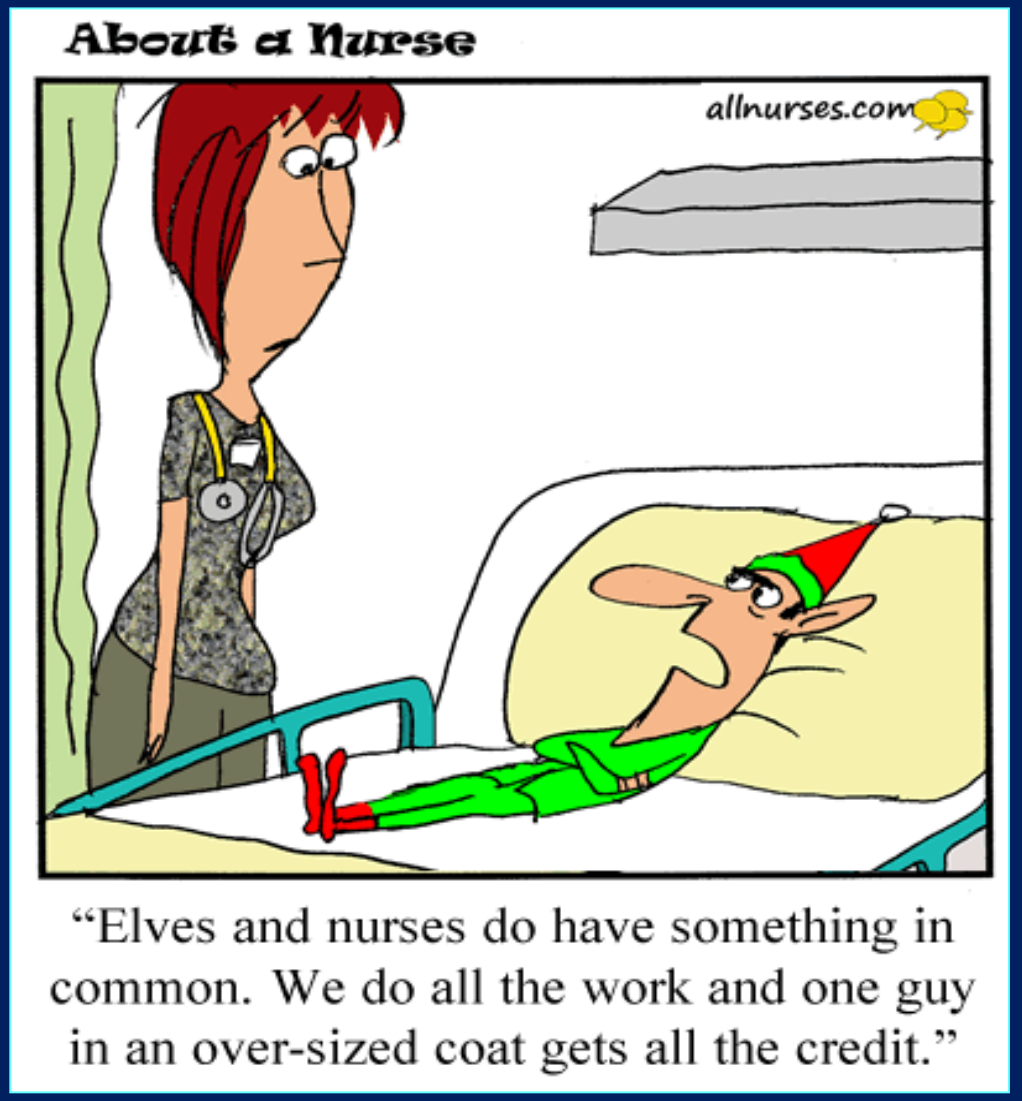
- Making notes in MDT meeting
  - Ensuring full follow up
- Maintaining DSD database
- Consent forms for research
- Maintaining referrals folder and spreadsheets
- Arranging admissions
- Creating pathways, protocols and information sheets

- Teaching
- Presenting / lecturing

- There for the family on the day
- Support when discharged
- Liaison with Psychology – support group days

In conclusion

- Complex but quick overview of the DSD service in London
- Brief explanation of what a DSD is
  - Types of referrals we receive
  - MDT management
- Clinical Nurse Specialist role
  - Liaison
  - Organisation
    - Specifics of the role
- Future for more advanced nursing roles?
  - Further training
  - Principle point of contact for new DSD referrals



Thank you

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