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

Kate Davies RN (Child), Dip HE, BSc (Hons), MSc
Clinical Nurse Specialist in Paediatric Endocrinology
Great Ormond Street Hospital for Children NHS Trust

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
  - CdG
  - 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 – Over 300 languages spoken

Great Ormond Street Hospital for Children

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

- Underground tube system oldest in the world
- 606 square miles
- 1.2 square miles City
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
      - CAH
    - Others
      - Hypoplasia

- 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
Revised nomenclature: Chicago Consensus 2006

<table>
<thead>
<tr>
<th>Previous</th>
<th>Revised</th>
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<tbody>
<tr>
<td>Intersex</td>
<td>Disorders of sex development (DSDs)</td>
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<td>Male pseudohermaphrodite</td>
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<td>Undervirilization of an XY male</td>
<td>46,XY DSD</td>
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<td>Undermasculinization of an XY male</td>
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<td>Female pseudohermaphrodite</td>
<td>46,XX DSD</td>
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<td>True hermaphrodite</td>
<td>Ovotesticular DSD</td>
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<td>XX male or XX sex reversal</td>
<td>46,XX testicular DSD</td>
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<td>XY sex reversal</td>
<td>46,XX complete gonadal dysgenesis</td>
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DSD MDT team at GOSH

- Biochemist
- Pathologist & Histopathologist
- Imaging & Urodynamics
- Endocrinologist
- Urologist
- Psychologist
- CNS
- Local Paediatrician
  - Surgeon / Urologist
  - Midwives / Nurses
- Clinical & Molecular genetics
- General Practitioner & Social Worker
- Community and Religious Leaders
- Ethicist
- Reproductive medicine
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**

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**GOSH DSD diagnosis statistics - 2014**
- 53 new referrals over one year
- Averaging 6 a month

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**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
- Aphallia
- Delayed / precocious puberty
- SOX2, SOX9
- Smith – Lemli – Opitz
- Mecham 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

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!
- Set a date and time for one day admission
Parents admissions leaflet

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, supplies, 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
  - [Usage with NICU / Respiratory ward

- 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, Hypoplastic, Klinefelter, AIS
  - Links to international DSD support groups
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
  - Apt for adrenal NLC 1/2 later
  - Liaise with local hospital, nursery, community nurses, ambulance services...
GP letter on day of discharge

- 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

- 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

Emergency hospital letter

Great Ormond Street Hospital for Children

Emergency Contact Details

- Name: [Name]
- Address: [Address]
- Phone: [Phone]
- Email: [Email]

In the event of an emergency, please contact:

- Great Ormond Street Hospital for Children
- Emergency Department
- Phone: [Emergency Phone]
- Email: [Emergency Email]

Sign here:

[Signature]

Date:

[Date]

Please note:

- This information is for emergency use only.
- If you require further assistance, please call the emergency services.
- This document is not intended for general information purposes.
Primary Care

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

Primary care – blood levels

- 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

- 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
- "Ehve and nurse do have something in common. We do all the work and one guy in an over-sized coat gets all the credit."

Thank you
References


