Anatomical and psychological challenges of management of children and adolescents with a disorder of sex development (DSD)

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What is our goal

- A young adult who is happy with his/ her gender assignment (gender identity and biological sex the same)
- An adult who is anatomically a perfect male or female, but unhappy with his/ her gender assignment is to me not a successful outcome

(This is a paediatricians point of view – the end results not available?)

What makes it difficult

- No patient is the same
- Complex etiology
- Different terminology (intersex, ambiquous genitaliae, true and pseudo hermaphrodites; masculinized/ virilized female and undervirilized male; DSD)
- The whole group has high prevalence of problems with sexual identification
- Excess androgens seems to imprint the foetal brain (eg female with congenital adrenal hyperplasia).
- Litigation due to “incorrect” decisions – even many years later
- Impossible to always get it correct

The different approaches - extremes

- Gender assignment seen as emergency in the neonatal period. Doctor makes a decision after a few basic tests, does the gender assignment and surgically correct the external genitalia to either a boy or mostly a girl
- Tests are done but no decision is made about sex. Child grows up as an “it” “gender neutral” and during teenage years decides what it wants to become. Surgery and hormone replacement is then started. This approach the result of litigation in USA against doctors and parents.

  Litigation because of psychological trauma due to no gender assignment?
- In-between. Patients managed individually. Do not implement protocols from other countries

In- between: Realistic approach

- First make a complete pathological diagnosis with facilities available. This is necessary for fertility prognosis and what to expect during puberty
- Explain to parents what the problem is and confirm with follow-up consultations.
- Delay decision about gender assignment – for months. Give parents a chance to observe their child. Assign gender early but delay surgery.
- Warn the parents that the management is going to be a long and delayed process and what the reasoning behind the delay is
- Keep contact with regular follow-up. Have seen many patients worked up completely and lost to follow-up. Seen many years later with catastrophic results.

Realistic approach 2

- Psychological support for child and parents
- Inform the child when appropriate age.
- Consult parents and other specialists to get consensus (surgeon, psychologist etc). Often not helpful because colleagues also unsure and nobody wants to commit. Parents usually fairly sure what they want.
- Paediatrician and surgeon must have a special interest in the condition
- Document everything. Files get lost. Patients get lost for follow-up. Some patients come from deep rural areas

Realistic approach 3

- Practical aspects should be kept in mind:
  - Anatomy of external genitalia
  - Problem of school attendance and social interaction of individual without clear gender assignment
  - Will hormonal replacement be available?
  - Will regular follow-up be possible?

Realistic approach 4

- Many patients tends more to one sex (parents raised, no doubt in their minds, external genitalia, doctor comfortable with the choice) – go ahead and treat preferably before school
- Some patients parents and the doctor are unsure or doctor not happy with parents choice.
  - Assign best fit gender but delay surgery
  - If forced to make a decision better to make a boy, because can be reversed. If the penis is removed one cannot go back.
  - There is an age limitation where testosterone will not enlarge the phallus anymore.

- Good luck with a difficult problem