

PUBERTY AND DISABILITIES

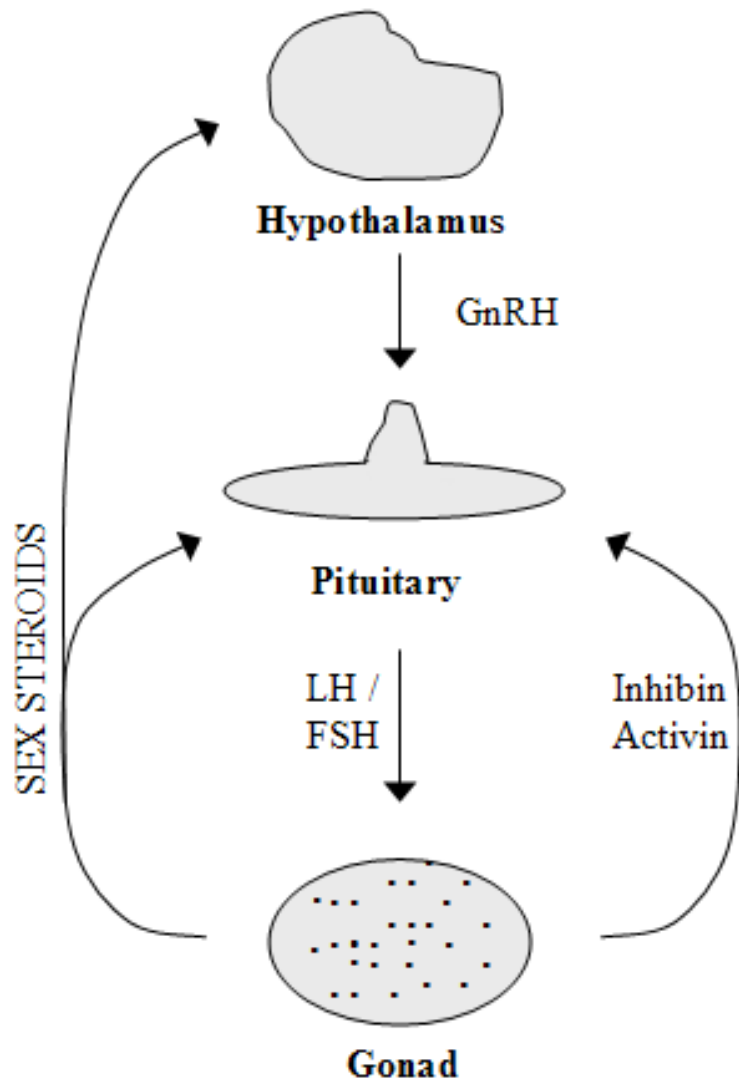
**Assunta Albanese
St George's Hospital
London**

Puberty

**Gonadal maturation with
acquisition of secondary sexual
characteristics and associated
growth spurt**



FERTILITY AND FINAL HEIGHT



Puberty

Average age of onset (UK):

- **11.0 yrs in girls**
- **11.2 yrs in boys**

Average age at menarche (USA):

- **African-American girls at 12.2 yrs**
 - **Caucasian girls at 12.9 yrs**
-



Changes during puberty

Events of Puberty

GIRLS

Thelarche

Pubarche

Menarche

Growth spurt

BOYS

- Testes & penis growth

- Pubarche

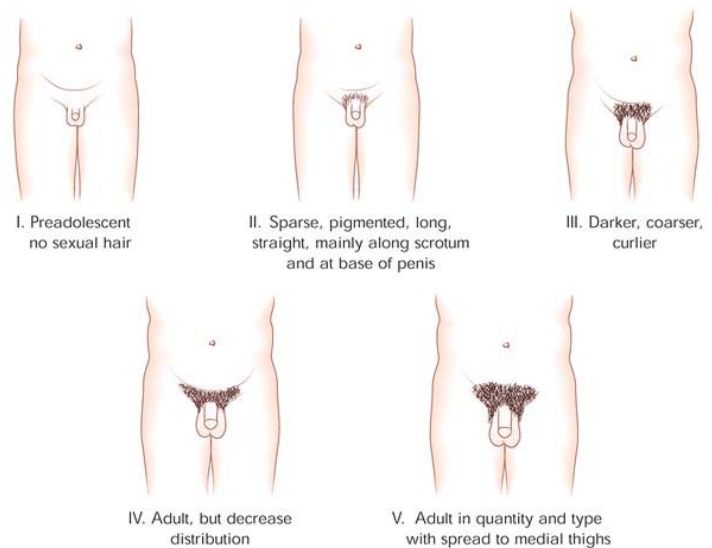
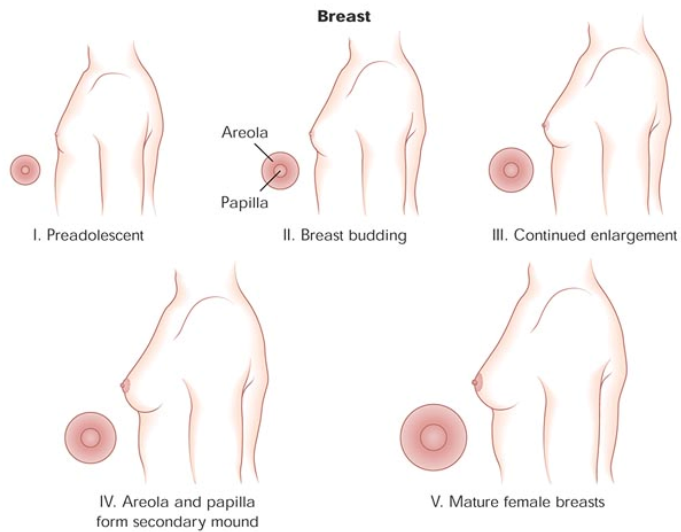
- Spermarche

- Body & Facial hair

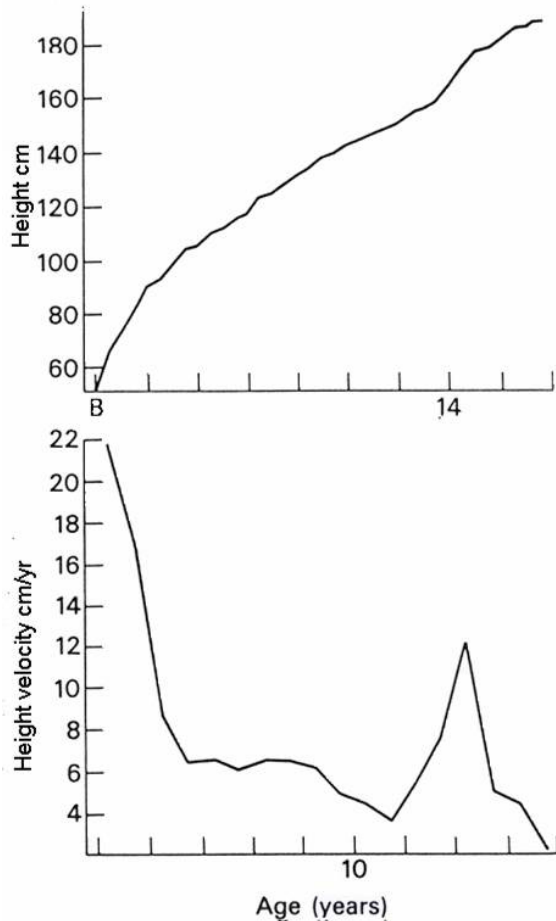
- Voice change

- Growth spurt

Pubertal Sexual Development



Pubertal Growth Spurt



- Pubertal growth accounts for about 20% of final adult height
- Pubertal total gain ~ 20-25 cm in females and 30 cm in males
- The average growth spurt lasts 24-36 months

Body Shape & Body Composition

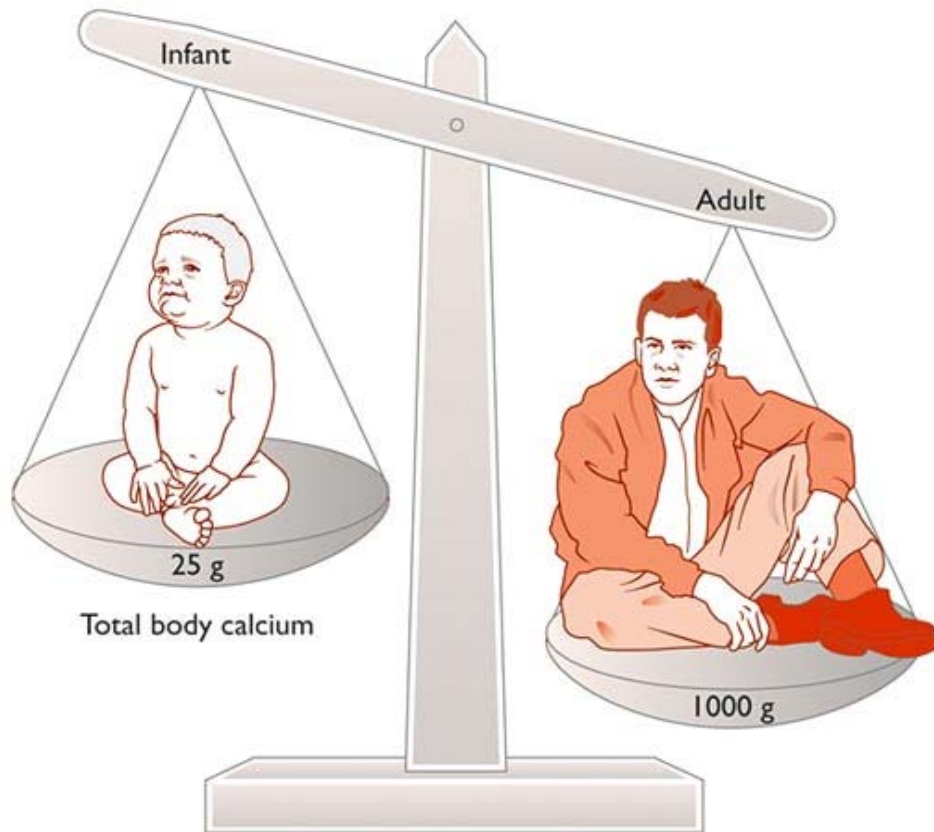
Male musculature vs female adiposity

- The average adult male has about 150% of the lean body mass of an average female, and about 50% of the body fat.

Bone growth

- Shoulder width and jaw in boys vs pelvis and hips in girls
-

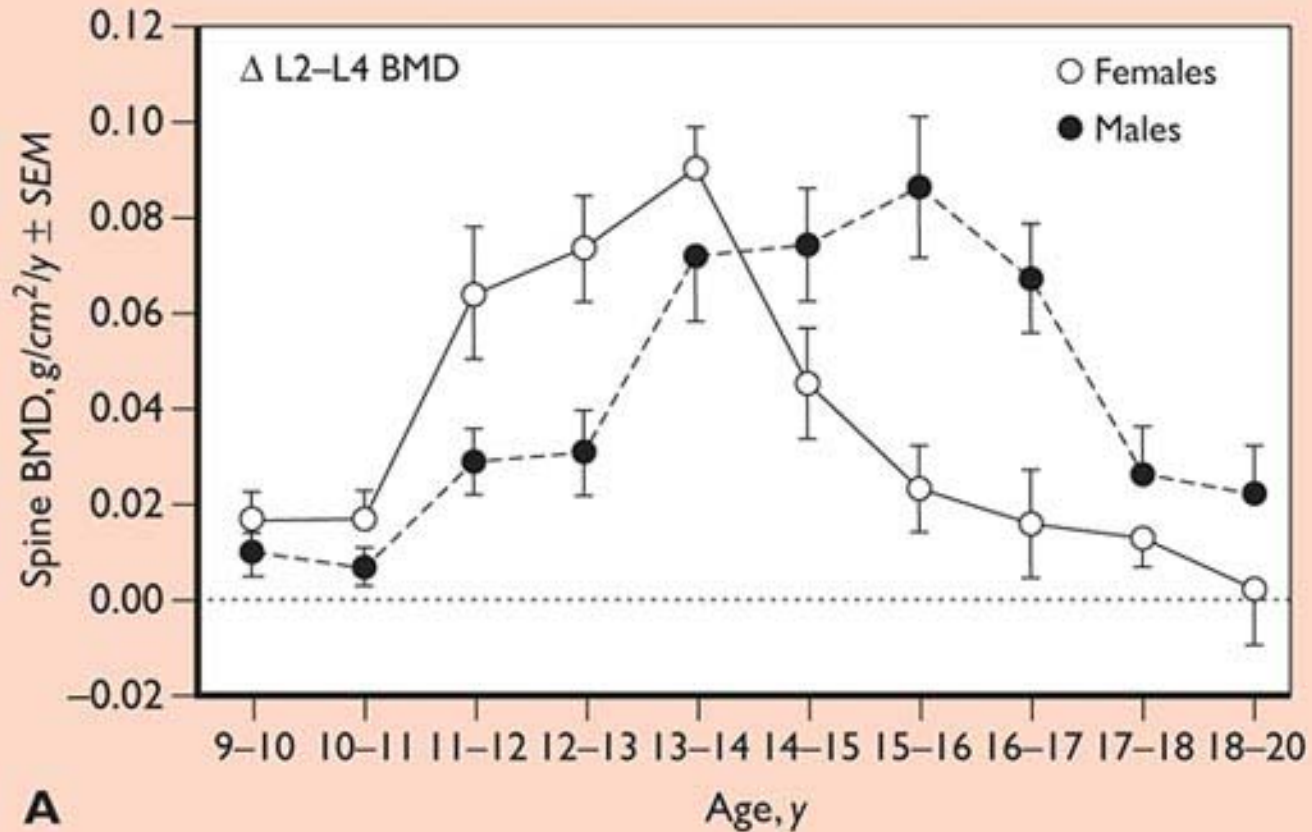
Bone Acquisition & Peak Bone Mass



The lifetime "bone bank"

- The increase in skeletal mass during puberty is critical
 - Peak bone mass is achieved by early adulthood
-

Bone Mineral Acquisition During Puberty



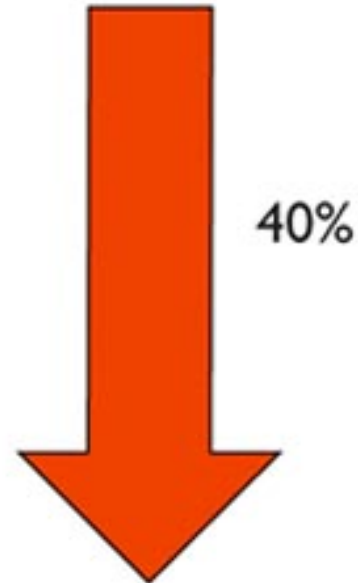
A

Peak Bone Mass and Osteoporosis Risk

Peak bone mass

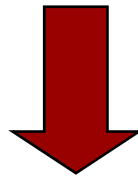


Osteoporosis risk

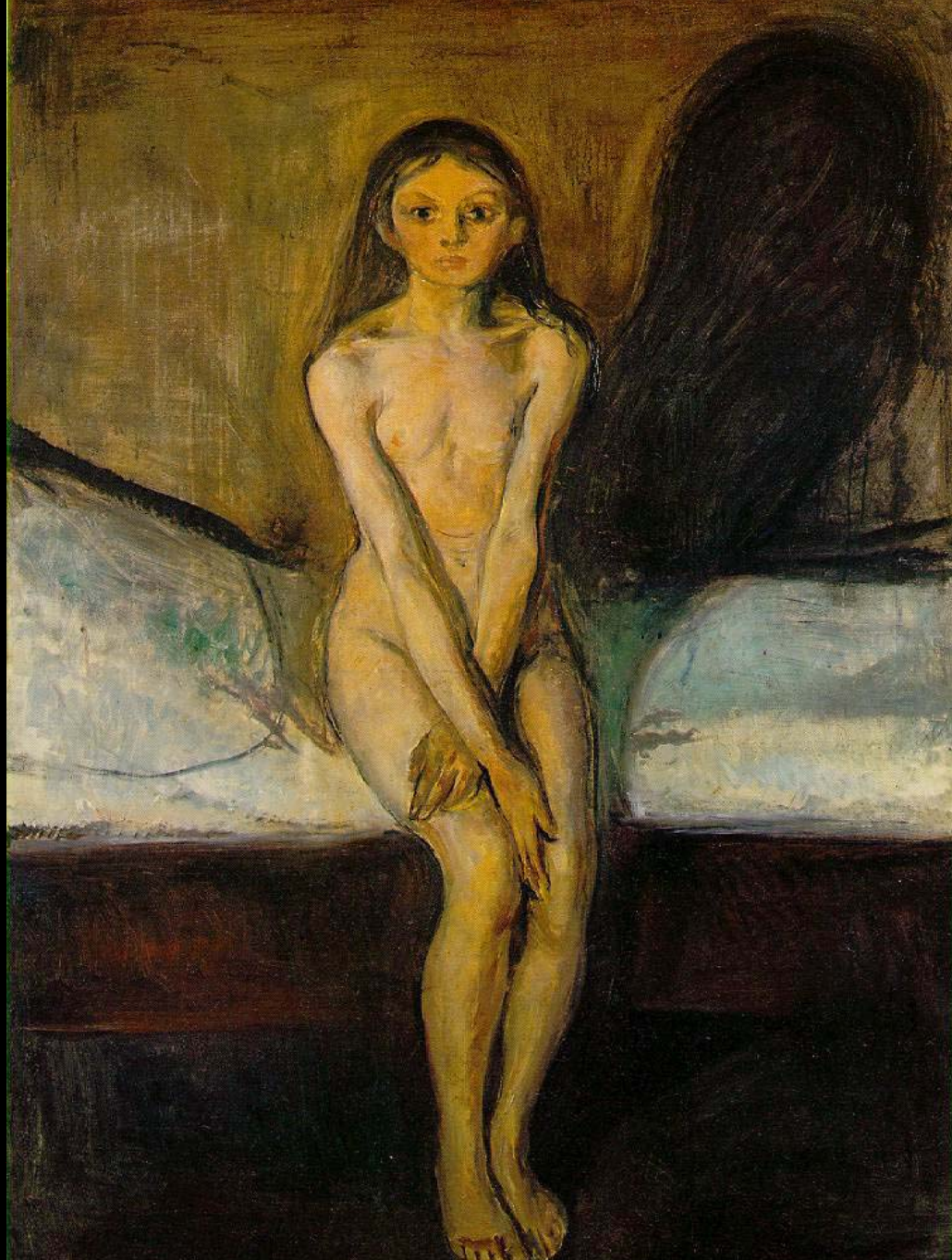


Puberty

**Gonadal maturation with acquisition
of secondary sexual characteristics,
associated growth spurt and
peak bone mass**



FERTILITY
FINAL HEIGHT
LIFETIME BONE BANK



Puberty and Disabilities:

Common requests for medical help

- Abnormal timing of puberty
 - Inability to cope with periods
 - Deterioration of epilepsy
 - Changes in behaviour
 - Feeling that patient should not deal with the “inconvenience”
 - Inappropriate sexual behaviour
 - Susceptibility to sexual abuse
 - Advice on safe sex/family planning
-

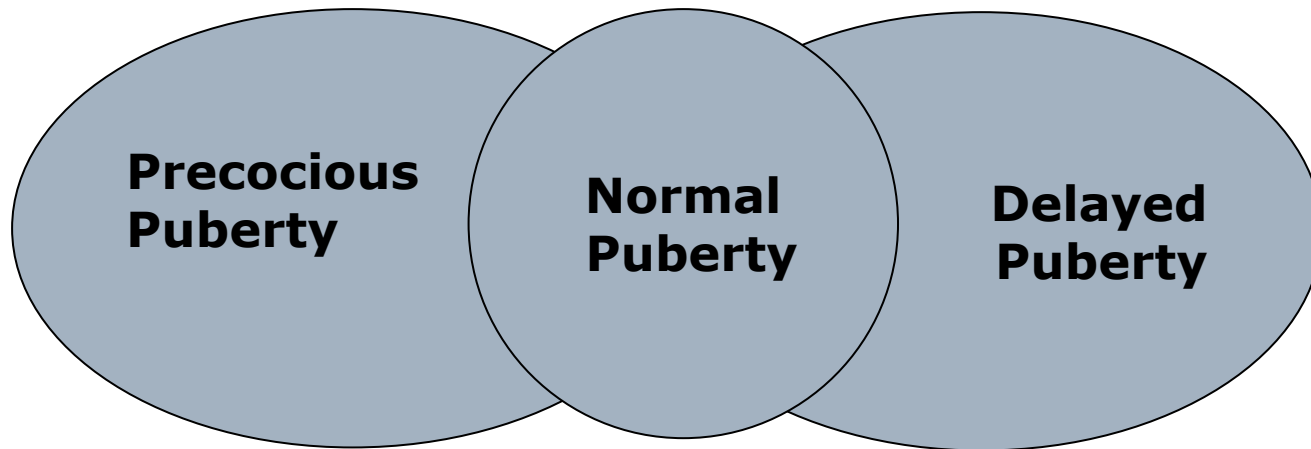
Clinical Problems Leading to a Paediatric Endocrinologist

- Hygiene issues 26%
- Precocious puberty 22%
- Fear of pregnancy 19%
- Fear of sexual abuse 16%
- Dysfunctional uterine bleeding 10%
- Specific sterilization request 5%
- Psychosocial concerns 1%
- "Inconvenience" osteoporosis risk 1%

Clinical Problems Leading to a Gynaecologist

<input type="checkbox"/> Need for advice or information	23.4%
<input type="checkbox"/> Menorrhagia	16.8%
<input type="checkbox"/> Need for contraception	11.2%
<input type="checkbox"/> Amenorrhea	9.3%
<input type="checkbox"/> Dysmenorrhea	8.4%
<input type="checkbox"/> Premenstrual mood or behavioural changes	5.6%
<input type="checkbox"/> Menorrhagia & irregular periods	5.6%
<input type="checkbox"/> Menorrhagia & need for contraception	5.6%
<input type="checkbox"/> Irregular menses	5.6%
<input type="checkbox"/> Irregular menses & need for contraception	3.7%
<input type="checkbox"/> Cyclical exacerbation of epilepsy	1.9%

Timing of Puberty



Precocious Puberty

Signs of pubertal maturation in:

- Girls under 8 yrs
- Boys under 9 yrs

In girls menarche under 9 yrs of age also indicate sexual precocity

First signs of pubertal maturation:

- Breast budding in girls
 - Increase in testicular volume in boys
-

Precocious / Early Puberty

- 20 times more frequent among children with neuro-developmental disability**
 - Puberty begins earlier but ends later in children with CP and moderate-severe motor impairment**
 - Effective treatment can be offered to arrest puberty**
-

Isolated Adrenarche

- ❑ Usually at around 6-8 years of age
 - ❑ Pubic/axillary hair, SOB, skin/hair greasiness, acne and changes in mood
 - ❑ Puberty at a normal time
 - ❑ Slight growth spurt with advanced BA
 - ❑ Final height prognosis not impaired
 - ❑ More common in neurologically impaired children
-

Delayed Puberty

- More common in boys than girls**
 - In specific conditions it can never occur due to a lack of gonadotrophins**
 - HRT to prevent osteoporosis**
 - Other risk factors for osteoporosis should be identified and corrected**
-

Coping with Periods

- Personal hygiene**
 - Excessive pain “she can't tell when it hurts or how it hurts”**
 - Heavy, frequent menstrual bleedings**
 - Effect of menses on routine & activities**
-

Coping with Periods

Mental Health Issues

- ❑ **Anxiety, mood changes and cyclical aggression**
- ❑ **Phobias: Fear of blood/ Refusal of wearing sanitary towels**
- ❑ **Acute exacerbation of psychiatric disorders**

Deterioration in Epilepsy Control

Consensus Statement: Menstrual and Contraceptive Management in Women with Intellectual Disabilities

The Australian Society of Adolescent Gynaecology Working Party

- Women with intellectual disabilities have the same right to the full range of management options as other women, tailored to their specific needs**
- Treatment options recommended should be the least restrictive and always in the best interest**

Consensus Statement: Menstrual and Contraceptive Management in Women with Intellectual Disabilities

The Australian Society of Adolescent Gynaecology Working Party

- ❑ Amenorrhea can generally be achieved by reversible ***less invasive*** methods than hysterectomy
 - ❑ Hysterectomy ***will not solve*** peri-menstrual behaviours or other cyclic problems
 - ❑ Many parents are concerned by sexuality and risk of abuse. Contraception, while preventing unwanted pregnancies, ***will not address*** either of these issues
-

Puberty and Disability

Women with disabilities may suffer endocrinologic disturbances such as:

- **Obesity**
 - **PCOS**
 - **Brittle diabetes**
 - **Hyperprolactinaemia**
 - **Thyroid problems**
-

Management of Periods

- ❑ Understand the reasons why help is requested
 - ❑ Good documentation
 - ❑ Awareness of “what is being asked for and what it is hoped can be achieved”
 - ❑ Frank discussion on “advantages” and “side effects” of interventions
-

Management of Periods: Issues for Consideration

- ❑ Level of functioning
 - ❑ Behavioural issues
 - ❑ Hand skills, capacity and practicalities of menstrual care
 - ❑ Acceptability of injections or oral drugs
 - ❑ Complicating medical factors, risk for osteoporosis, DVT
 - ❑ Interaction between antiepileptic / other drugs and oestro-progestin
-

Interventions

- Education
 - Psychotherapy
 - Medical
 - Surgical
-

Puberty and Disabilities

- Appropriate education and support
 - Children with higher support needs may require more basic information such as:
 - Differences between males and females
 - The changes of puberty
 - Appropriate or inappropriate touch
 - How to behave in different social situations
 - Irregular bleedings are common in all girls after menarche
-

Intervention

Behaviour

- ❑ Routine
- ❑ Rewards for appropriate behaviour

Social

- ❑ Education with peer groups; understanding/accepting

Psychotherapy

- ❑ Social stories
 - ❑ Modified cognitive behavioural therapy
-

Medical Therapy: Dysmenorrhoea

- Pain or behaviour changes thought to reflect pain can be initially managed with non-steroidal anti-inflammatory drugs (NSAIDs)
 - **Predicting to be in pain for the first 2 days of each cycle**
-

Hormonal interventions

- Depo-Provera**
 - OCP or Continuous OCP**
 - Mirena**
 - GnRH analogues**
-

Depo-Provera

- IM injection 12 – weekly
 - Usually causes amenorrhoea – spotting in up to 50%
 - Potential side effects:
 - Mood and behavioural changes
 - Weight gain
 - Pain from injection
 - Fatigue
-

Depo-Provera

- ❑ Well established link with decreased BMD in adults and adolescents
 - ❑ Critical time for BMD acquisition
 - ❑ CSM warning, FDA black box
 - ❑ BMD probably regained on discontinuation
 - ❑ Vit D and Ca supplementation
 - ❑ No data on fracture risk / osteoporosis
 - ❑ No data on immobile population
-

Continuous OCP

- ❑ Extended use of COCP allows less frequent periods with more control
 - ❑ Oral administration
 - ❑ Anticonvulsants may reduce efficacy
 - ❑ Thromboembolic risk esp if immobile
-

Mirena ®

- ❑ Intrauterine system
 - ❑ Progesterone reservoir releases over 5 years
 - ❑ Used for primary menorrhagia
 - ❑ Decreases menstrual loss by 90%
 - ❑ Concerns over communication of complications
-

GnRH analogues

- ❑ Injectable 4 weekly / 12 weekly
 - ❑ Hypo-oestrogenic state
 - ❑ Expensive
 - ❑ BMD loss
 - ❑ Add-back HRT / Tibolone protective in adults
 - ❑ No data on use in adolescents
-

Surgery

- Endometrial ablation
 - Limited experience
 - Decrease menstrual losses or amenorrhea in 60%
 - Needs repeat procedures
 - May cause sterility
 - Hysterectomy
 - Consent issues very difficult
-

Conclusions 1

- Planning ahead...**
 - Educational/ psychological support to child and family**
 - Pharmacological interventions are available and should be tailored to the patient's needs**
-

Conclusions 2

- **Long term profound “hypo-oestrogenism” should be avoided**
 - **BMD in at-risk patients needs to be assessed and monitored serially**
-

Conclusions 3

- ❑ **Current treatment options have limitations**
 - ❑ **No long term medical strategies exists to completely suppress periods without the possibility of adverse consequences**
-