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Friday, August 09, 2019

(Room 5)

16:30 - 18:30 WS #54: Paediatric Forum (120mins, not repeated)

Common Endocrine Problems

Dr Martin de Bock PhD

Contents

- Diabetes
- Growth
- Puberty

Diabetes concerning

- DKA at presentation occurs in 40% of children according to international averages.
- Canterbury 2018, that figure was 70%
- Multifactorial BUT
  - Repeat presentations to GPs
  - Outpatient bloods and referrals instead of acute diagnosis and admission
  - 38 children diagnosed in Canterbury in 2018.

Different presentations of diabetes mellitus

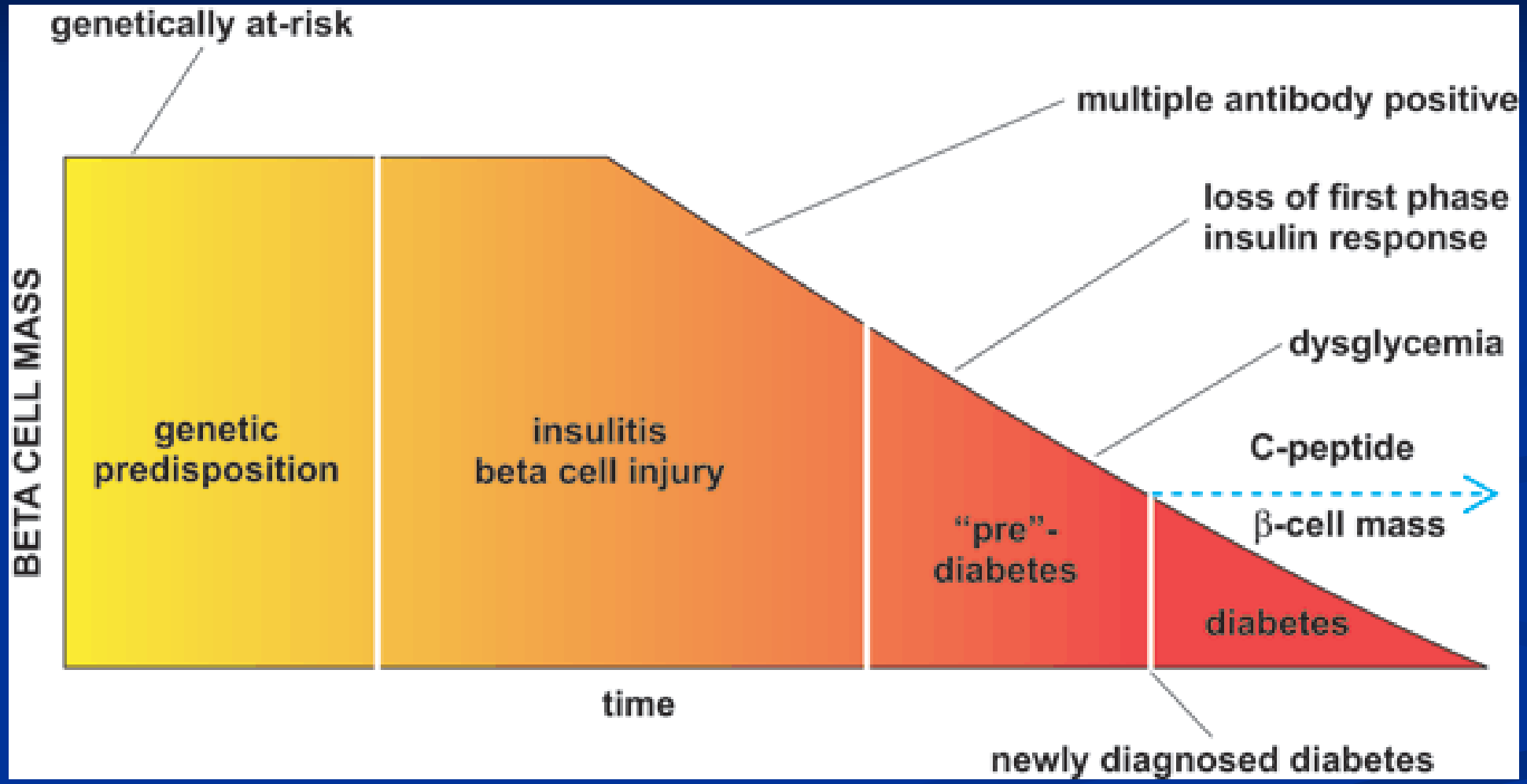
- Can present throughout infancy, childhood and adolescence
- POLYDIPSIA and POLYURIA, weight loss – cardinal features of diagnosis (need to ask)
- Diabetic ketoacidosis can present with abdominal pain and vomiting, heavy or rapid breathing, on background of polydipsia and polyuria

Risk of Type 1 Diabetes compared with other chronic childhood diseases

Disease	Rate/100,000
Type 1 Diabetes	24
Cancer (< 15yr) Overall	15.6
Leukemia	5.3
Brain	3.6
Cystic Fibrosis	3.1
J.C.A.	3.0
Nephrotic Syndrome	1.3

Australian Paediatric Cancer registry, 2009

ABS 2010



>90% beta cell function loss: Type 1 diabetes

Diagnosis

- Typical symptoms AND
- Urine positive for ketones and glucose
- Blood glucose: random > 11 mmol/L

  fasting > 7 mmol/L

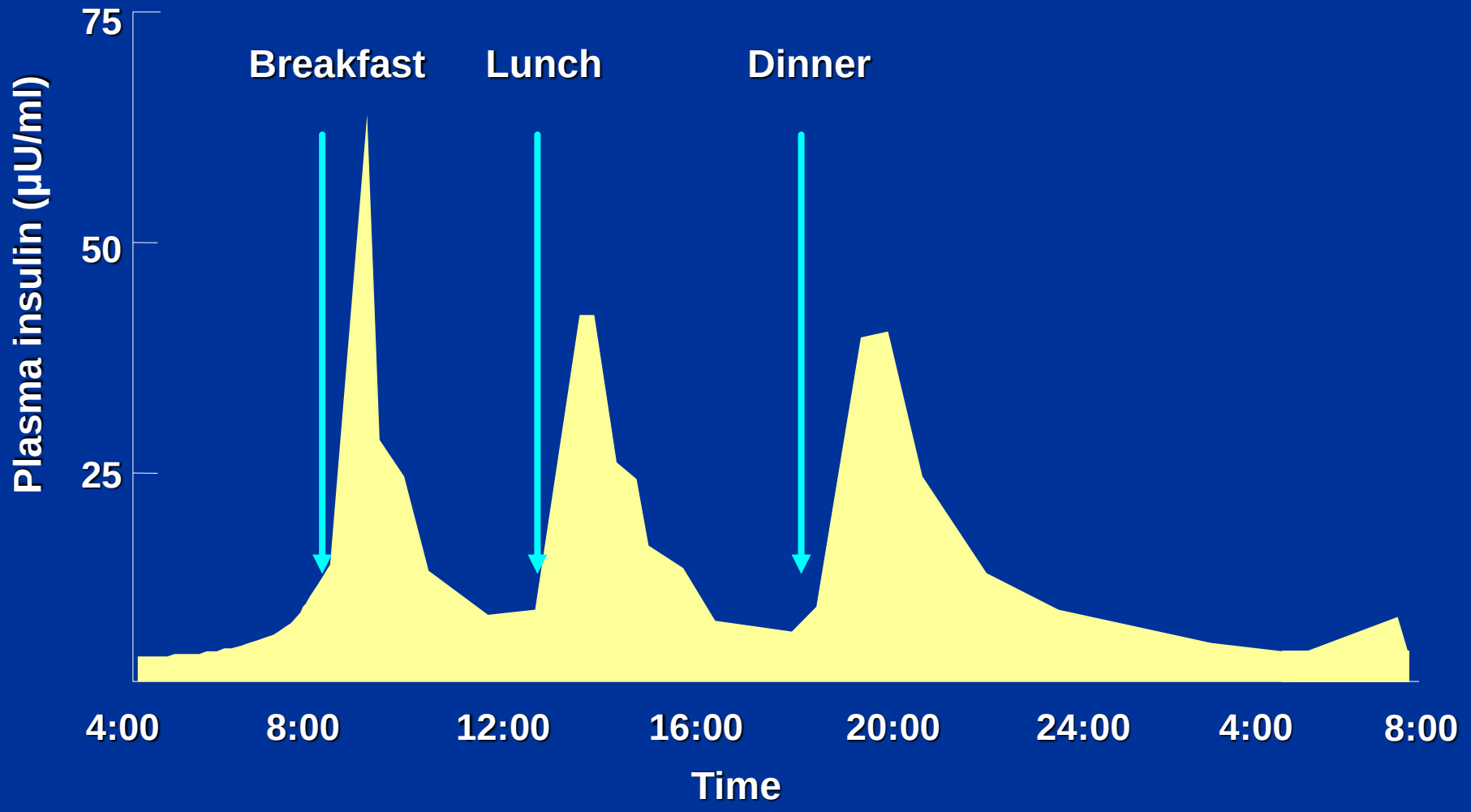
OGTT NOT INDICATED

Urgent referral is necessary

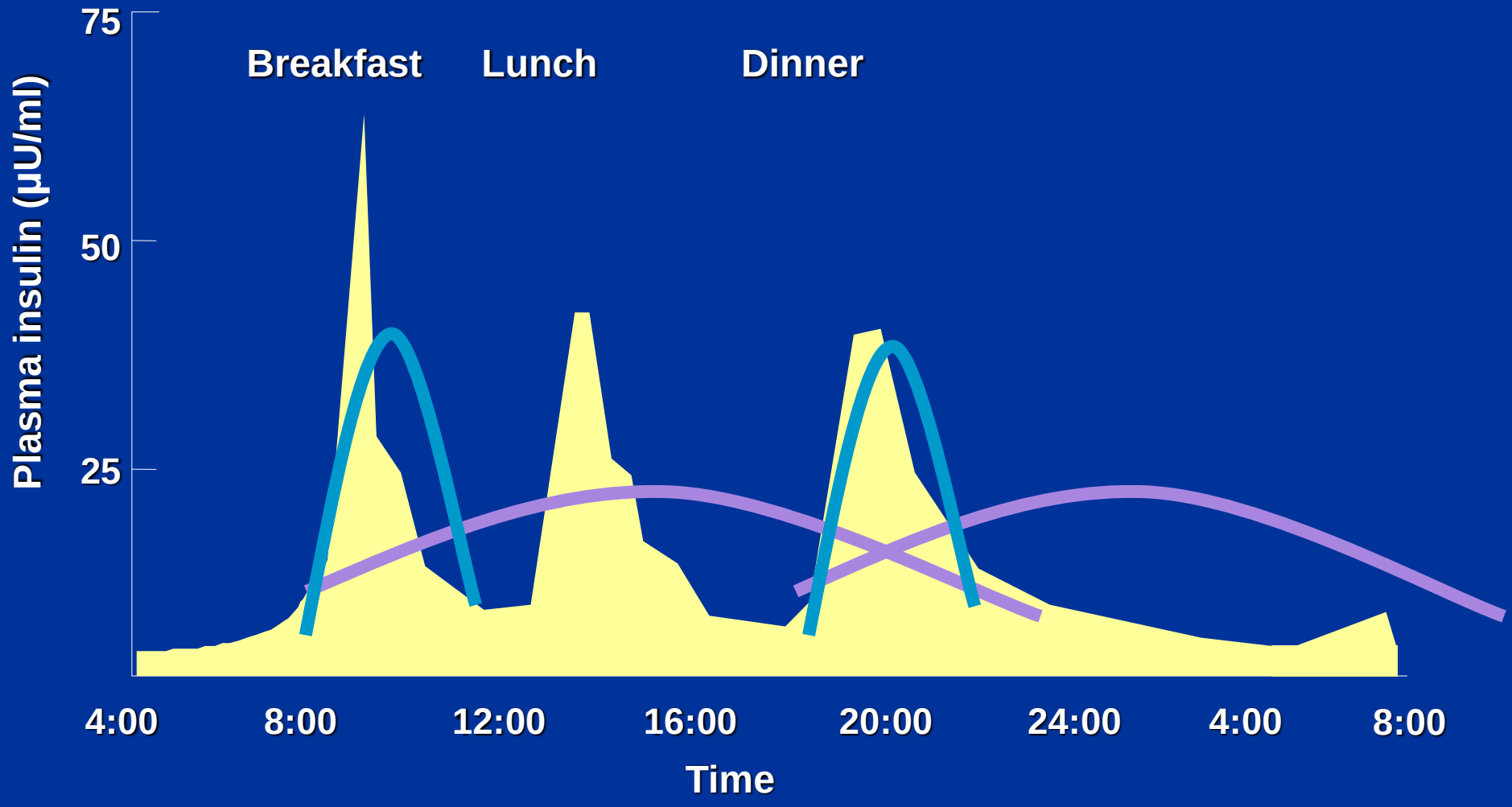
Outpatient Insulin Therapy

- Try to mimic physiological insulin secretion
- - illustrated in the following slides:
  - Physiological profile
  - BD insulin regimen
  - MDI
  - Insulin pump

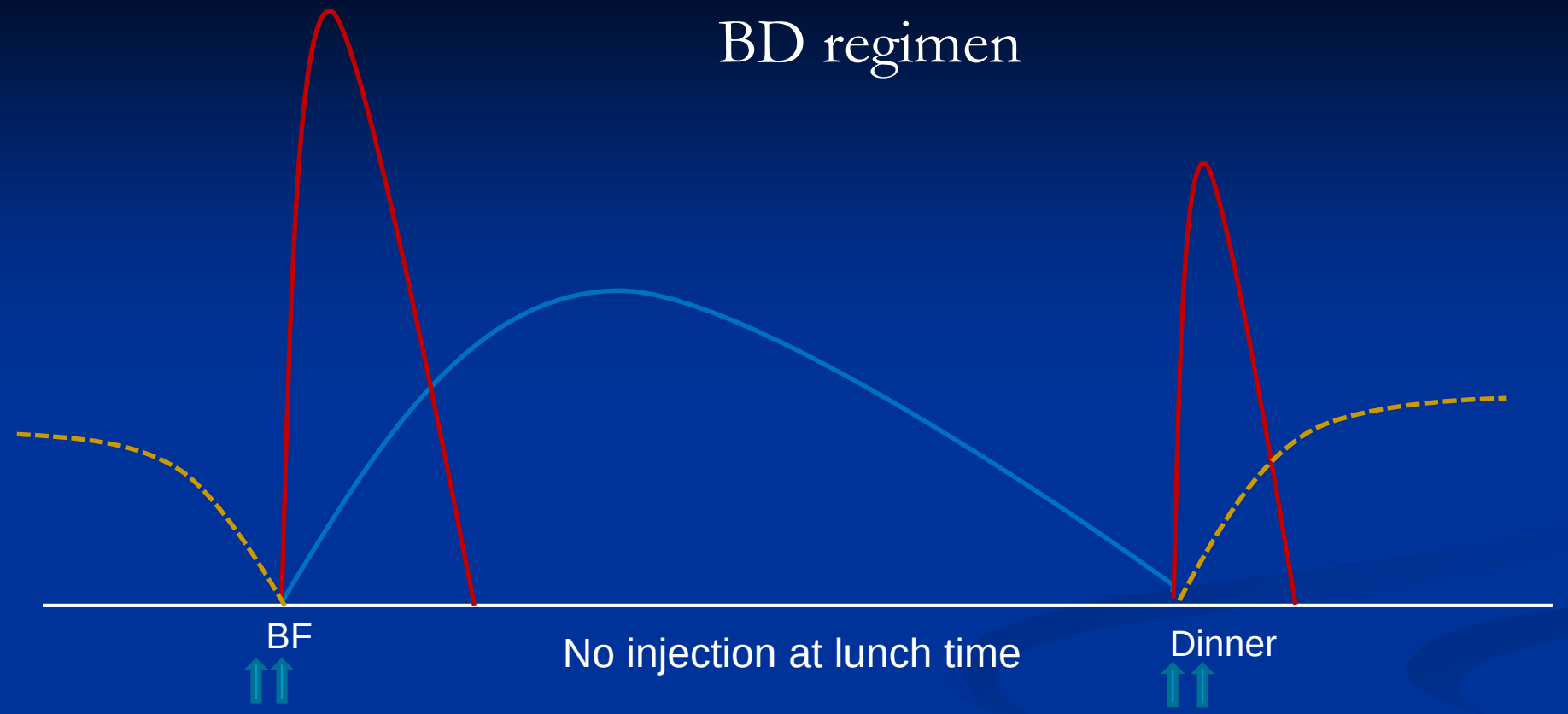
Physiological Serum Insulin Secretion Profile



BD regimen – mixture of quick and intermediate-acting insulins given twice daily

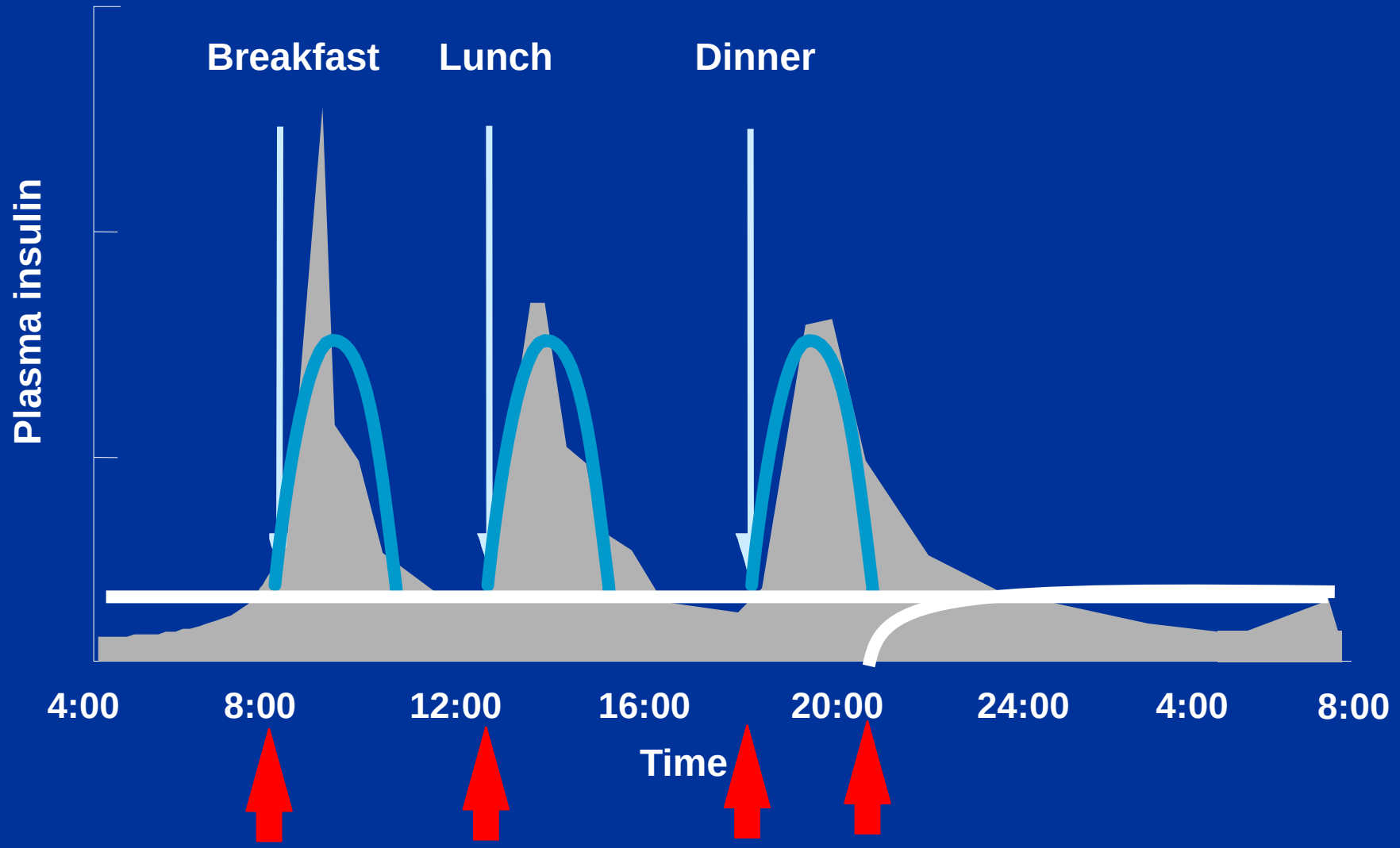


BD regimen

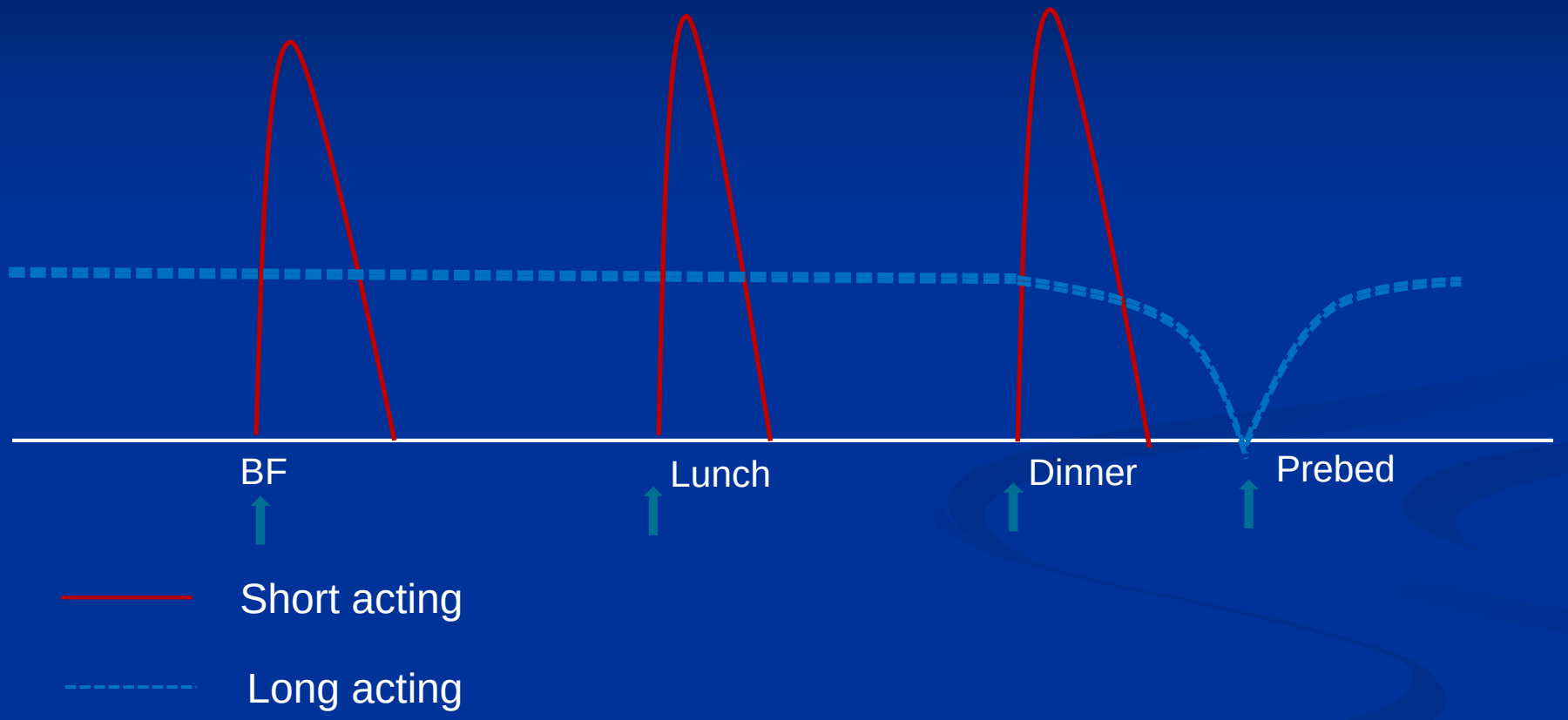

BF
No injection at lunch time
Dinner
Short acting
Intermediate acting
Long acting

Basal/Bolus (MDI) – quick insulin

before meals and long before bed)



Multiple daily insulin (MDI) regimen



Insulin pump :

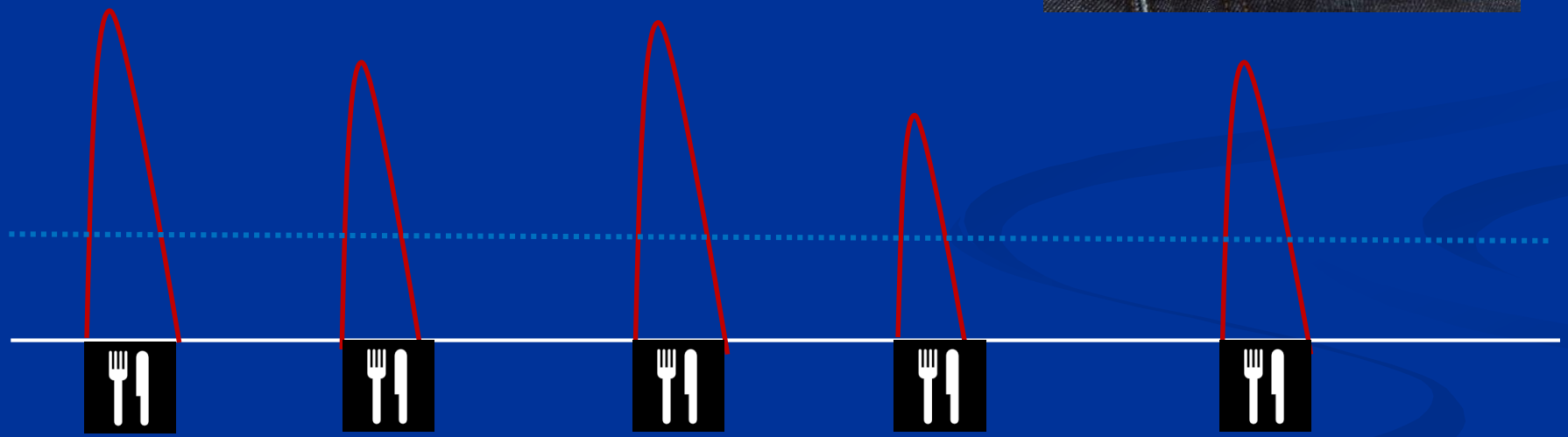
continuous infusion of rapid acting insulin

Insulin pump

Bolus

Basal

Short acting insulin



New Technologies

- Flash glucose monitoring
  - No calibrations
  - $100 a fortnight
  - Hopefully PHARMAC will fund

- Automated insulin delivery
  - Commercially available Australia, Europe, USA
  - Research occurring in NZ

GP role

- Consolidation of support
- Transition
- Other issues
  - Acne
  - Mental health
  - Prescriptions

Diabetes TAKE HOME MESSAGE

- Diagnose T1D in the clinic
  - Urine, or finger prick
  - An OGTT is not indicated
  - If you are ticking HbA1c on a form for a child, please check the BG and urine at that time, as you might need to send to hospital NOW

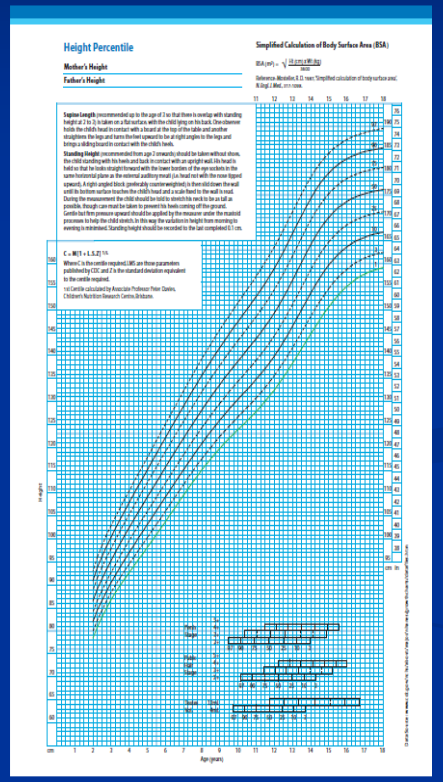
Growth and Puberty

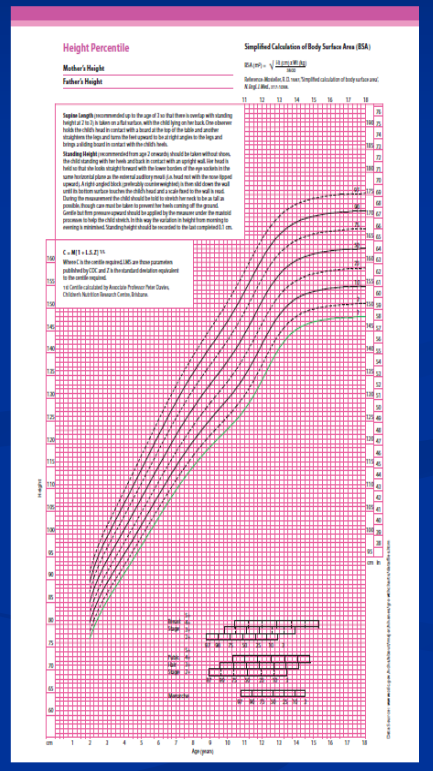
Growth monitoring: an essential part of child health surveillance

- Height and weight should be measured whenever child attends hospital/clinic – serial measurements important
- Stadiometer
- Plot on appropriate growth charts (height, weight, ht/wt velocity)

Growth Chart

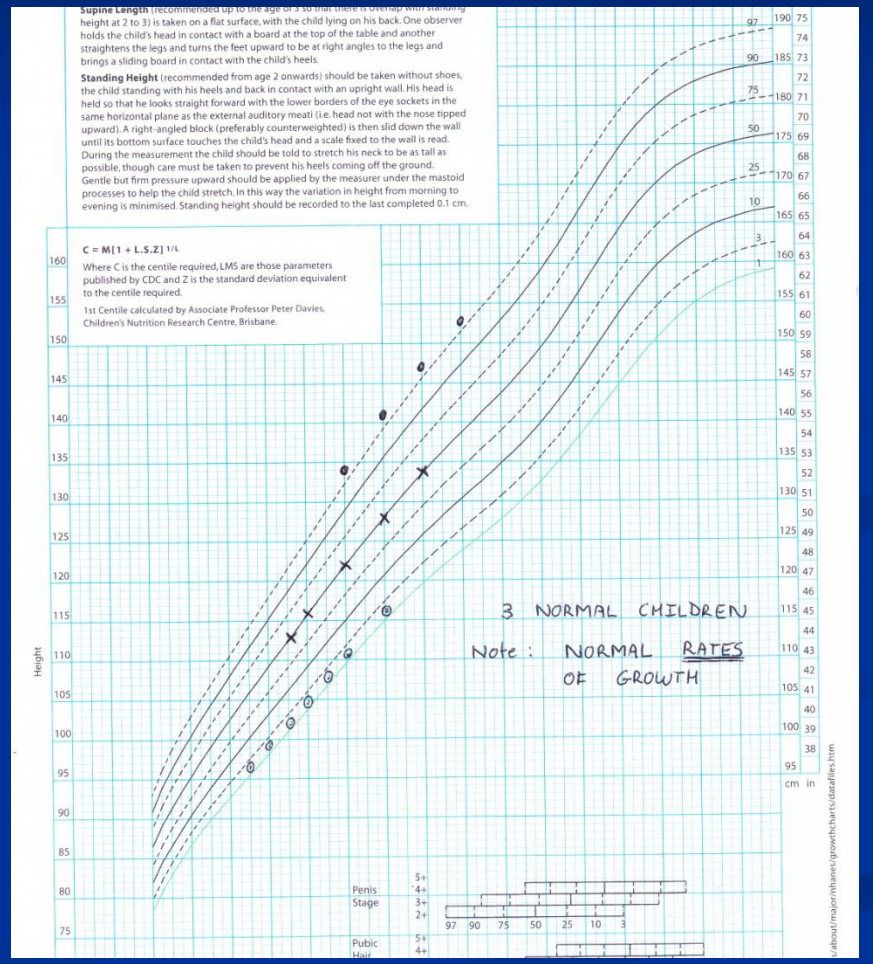
- Demonstrate the range of normal growth
- Percentile curves derived from the normal distribution (bell-shaped curve) of the growth data



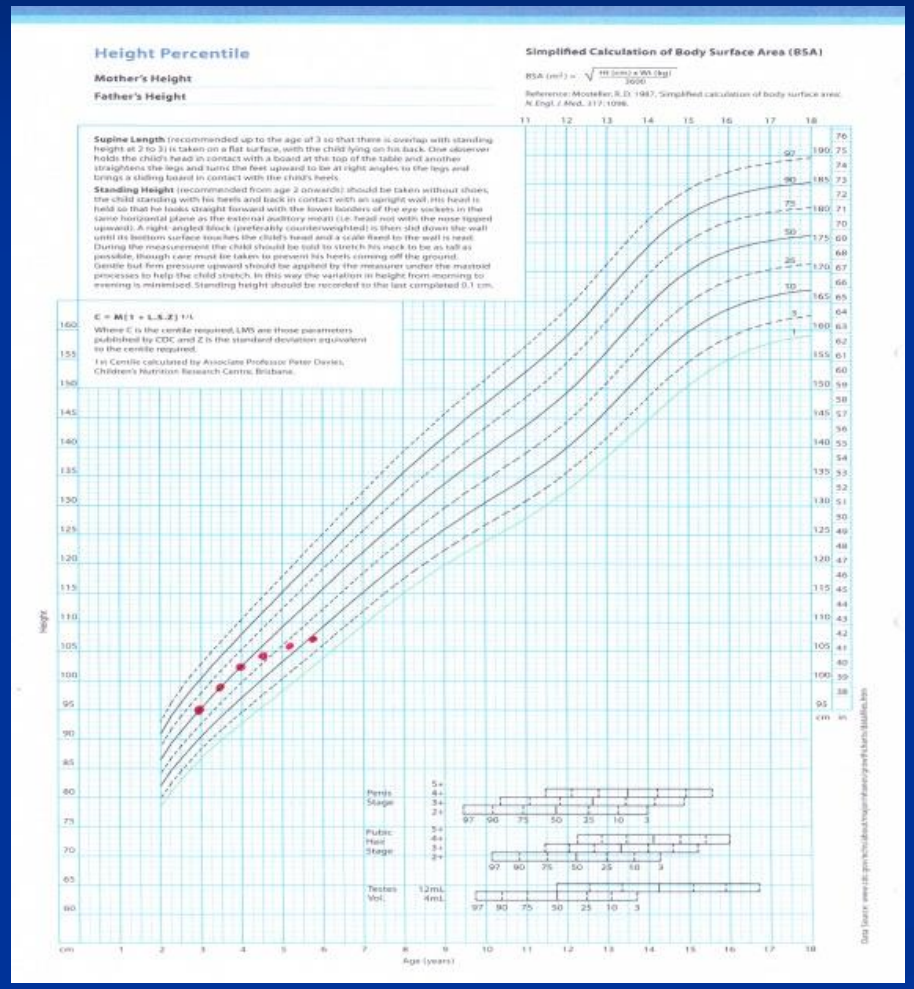


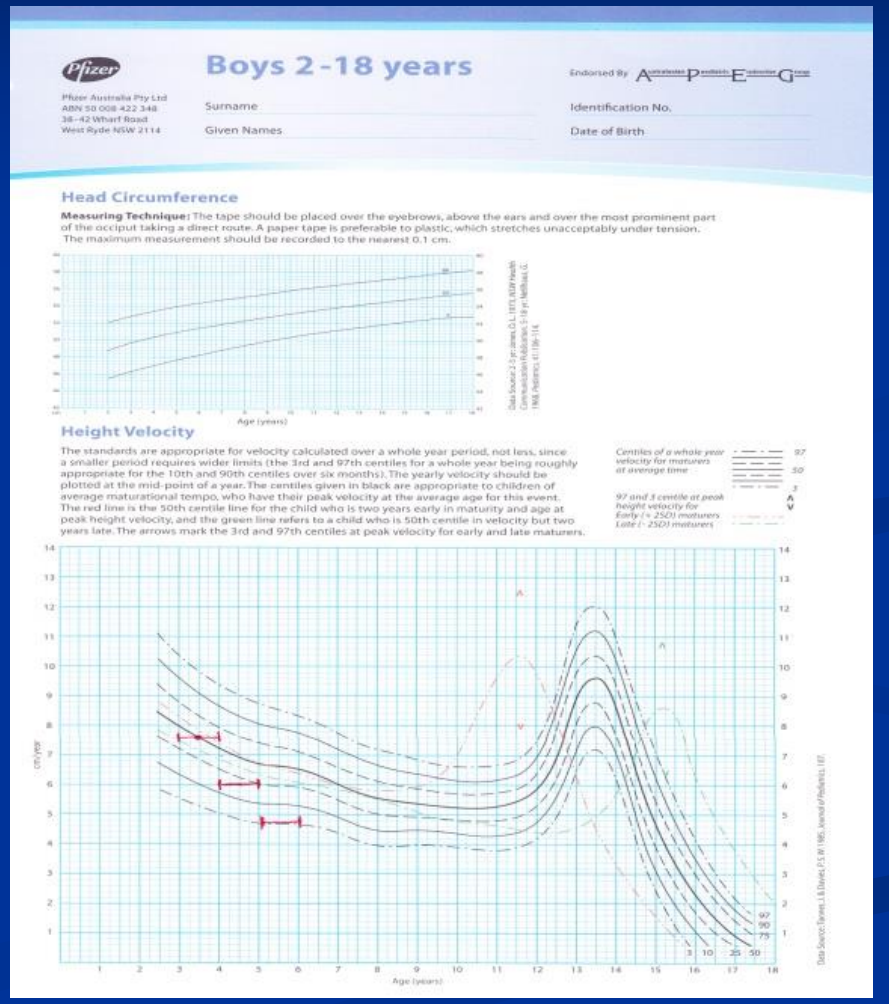
Growth chart

- Median is 50th percentile: 50% of children lie above and 50% below
- 3% of NORMAL children will be <3rd percentile and 3% above 97th
- 3rd and 97th percentiles approximately represent 2 SD from the mean



Growth velocity

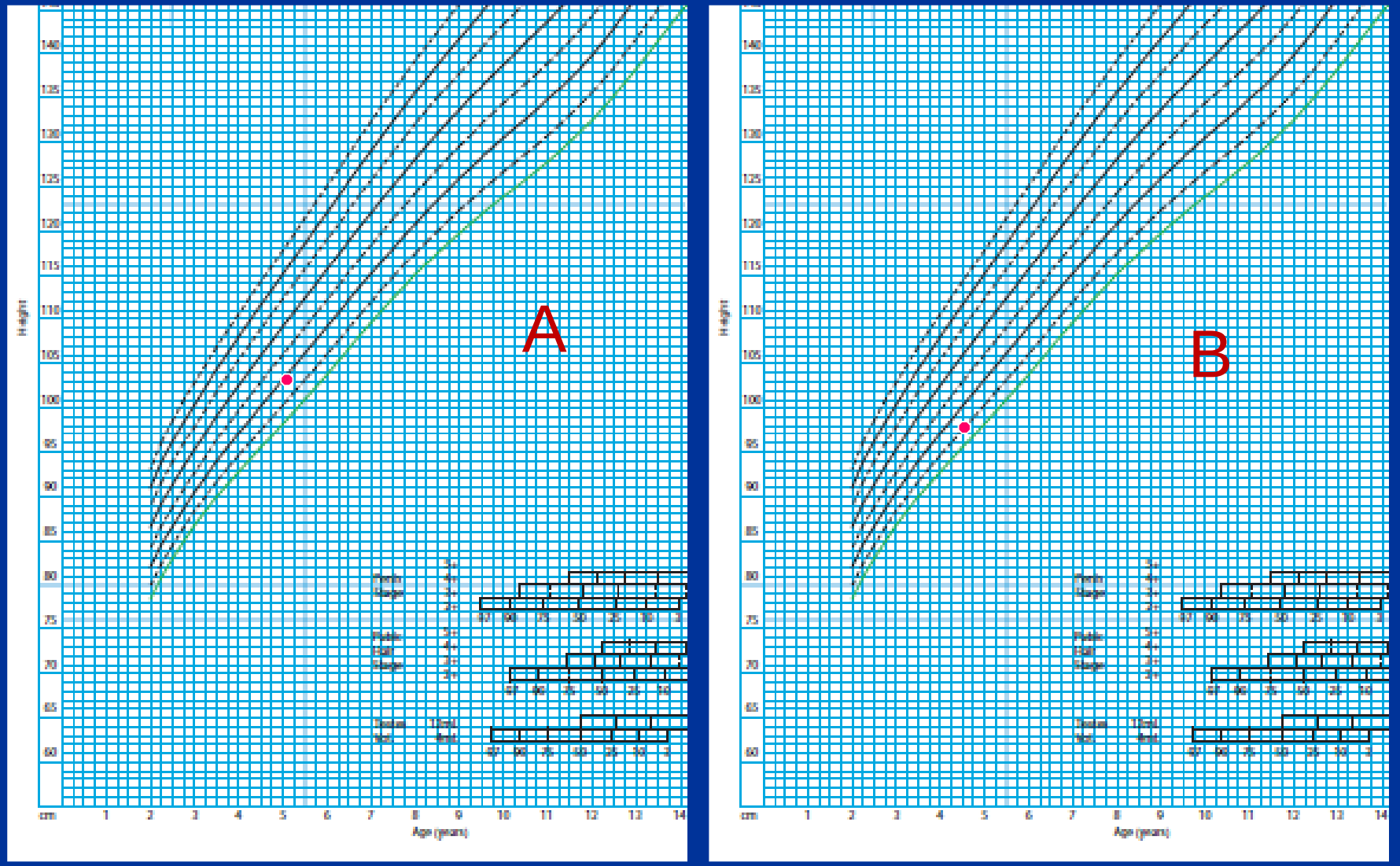




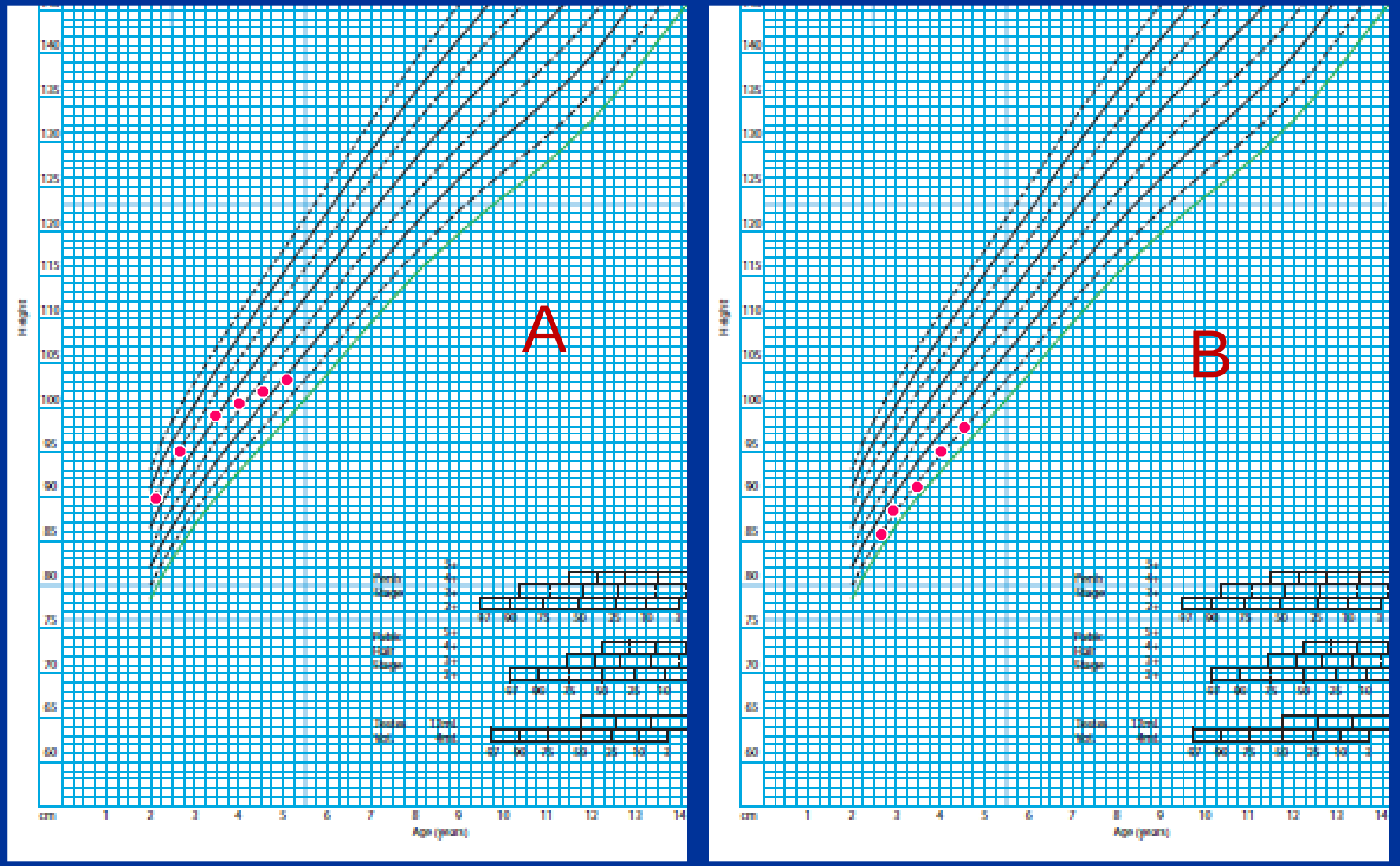
Growth rate (velocity)

- Much more important than single measurement or position on chart
- Based on serial measurements (3-6 monthly) over 1 year
- Most children tend to follow their centile – SIGNIFICANT DEVIATION IS IMPORTANT

Which child are you more worried about?



Which child are you more worried about?



Assessment of short stature

- Is he/she short? Chart on growth chart
- Is he/she growing slowly?

  - SERIAL measurements/growth trend

- What is the cause? Direct history, examination and investigations towards these causes

Causes of Short Stature

- VARIATIONS OF NORMAL
  - Genetic/familial short stature
    - Low genetic height potential, height tracks parallel to 3rd centile, growth velocity is normal, normally timed puberty
  - Constitutional delay in growth and puberty
    - "late bloomer"

Determining genetic potenetial *interest only!

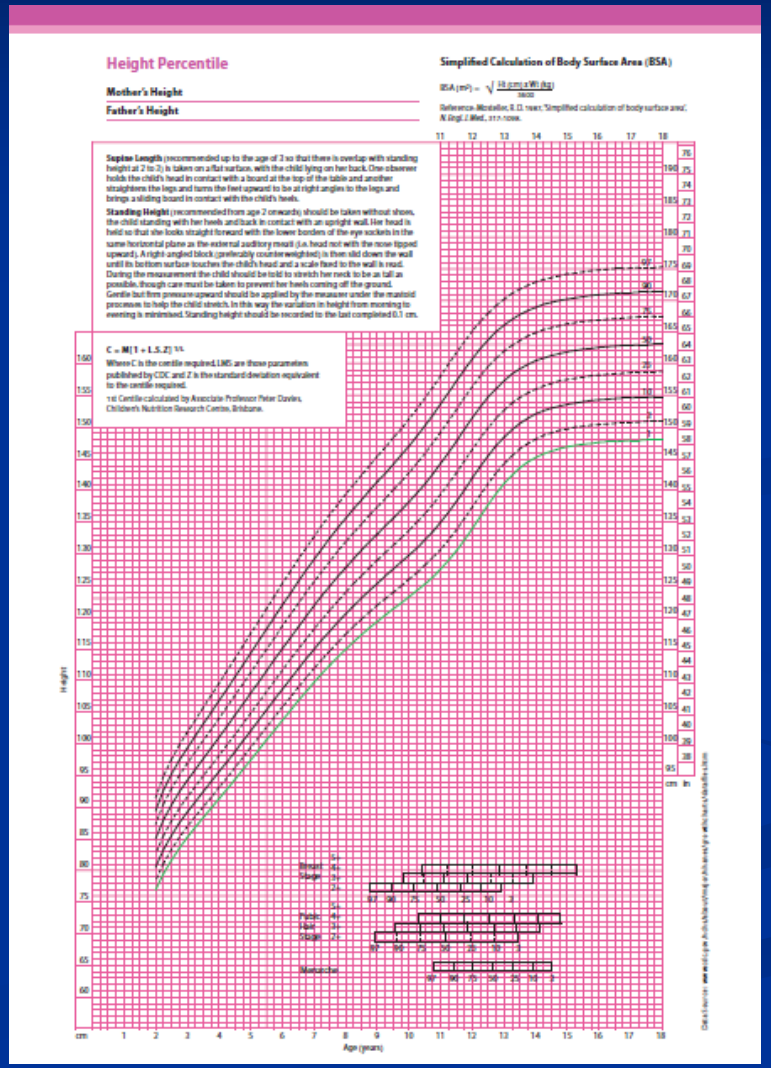
Boys:

- $\frac{\text{M Ht + F Ht +13 cm}}{2}$

Girls:

- $\frac{\text{M Ht + F Ht – 13 cm}}{2}$

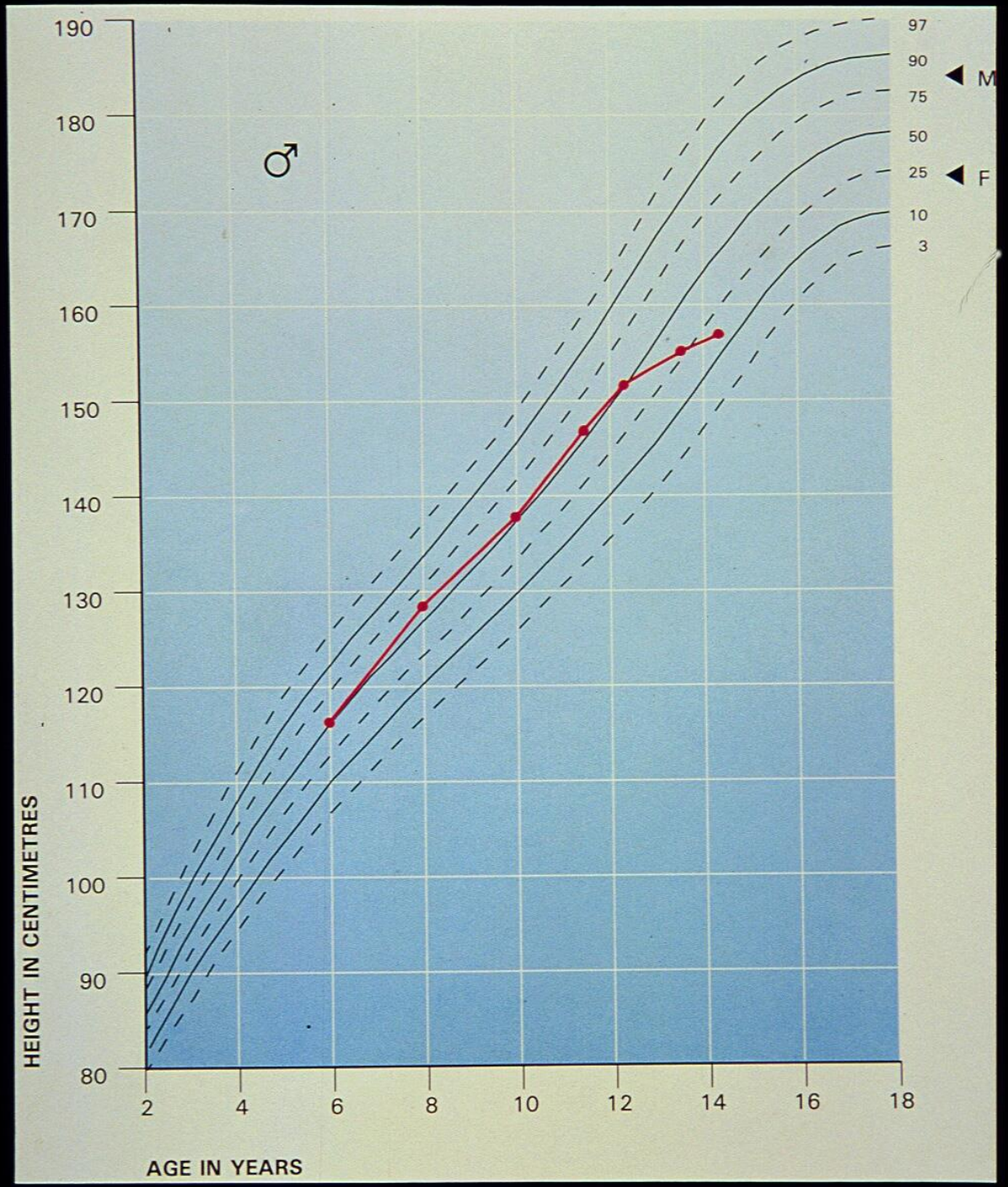
Target Range
±6.5 cm



- Pathological
  - SGA
  - Chronic illness/malnutrition/drugs
  - Chromosomal abnormality/syndrome
  - Psychosocial
  - Skeletal dysplasia
  - Endocrine (growth hormone deficiency, thyroid hormone deficiency, steroid excess, pubertal delay)

Short and lean

Short and fat

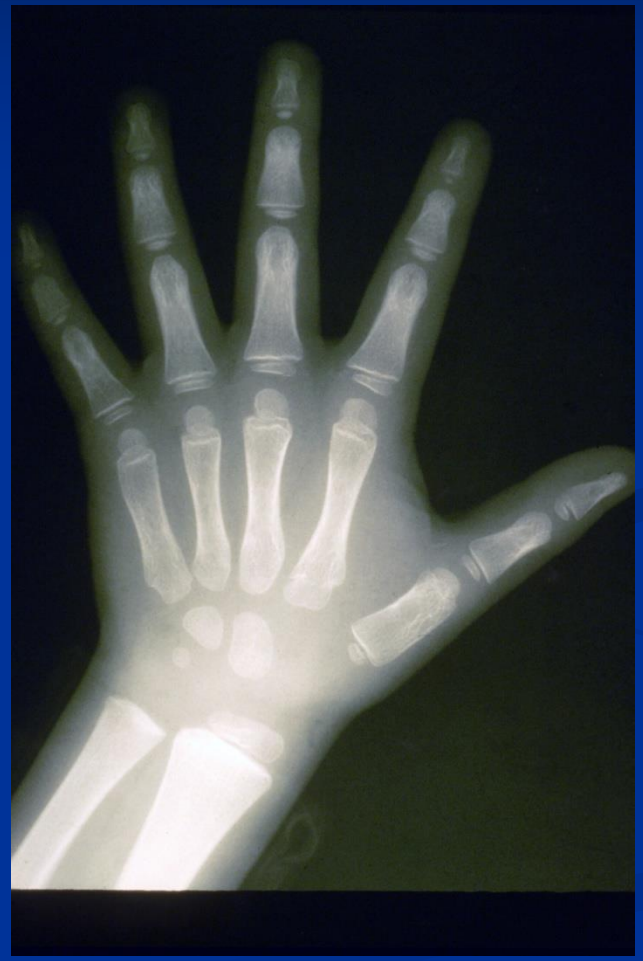

♂
190
180
170
160
150
140
130
120
110
100
90
80
HEIGHT IN CENTIMETRES
2
4
6
8
10
12
14
16
18
AGE IN YEARS
97
90
75
50
25
10
3
M
F

Constitutional delay of growth + puberty

- Common variation of normal growth but exclude pathological causes of slow growth
- More common in boys
- Usually family history of similar growth pattern
- Look younger than age, later puberty, grow for longer, final target height preserved, bone age delayed for chronological age*

Next slide

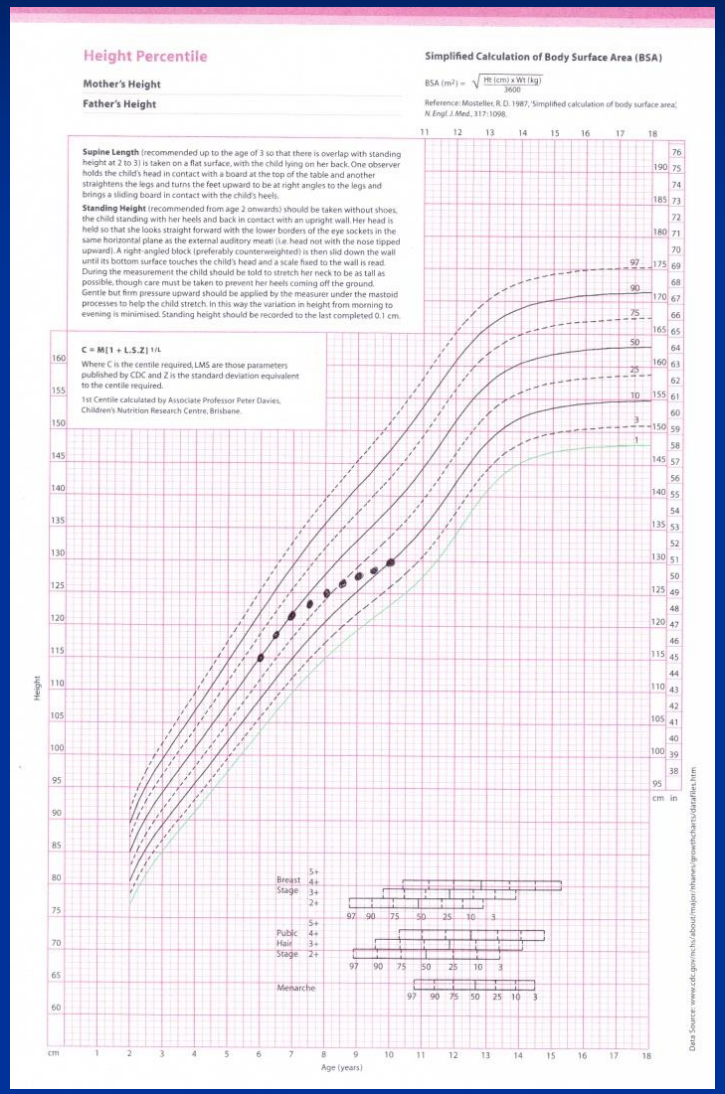
Bone age X-ray
(Left hand and wrist)

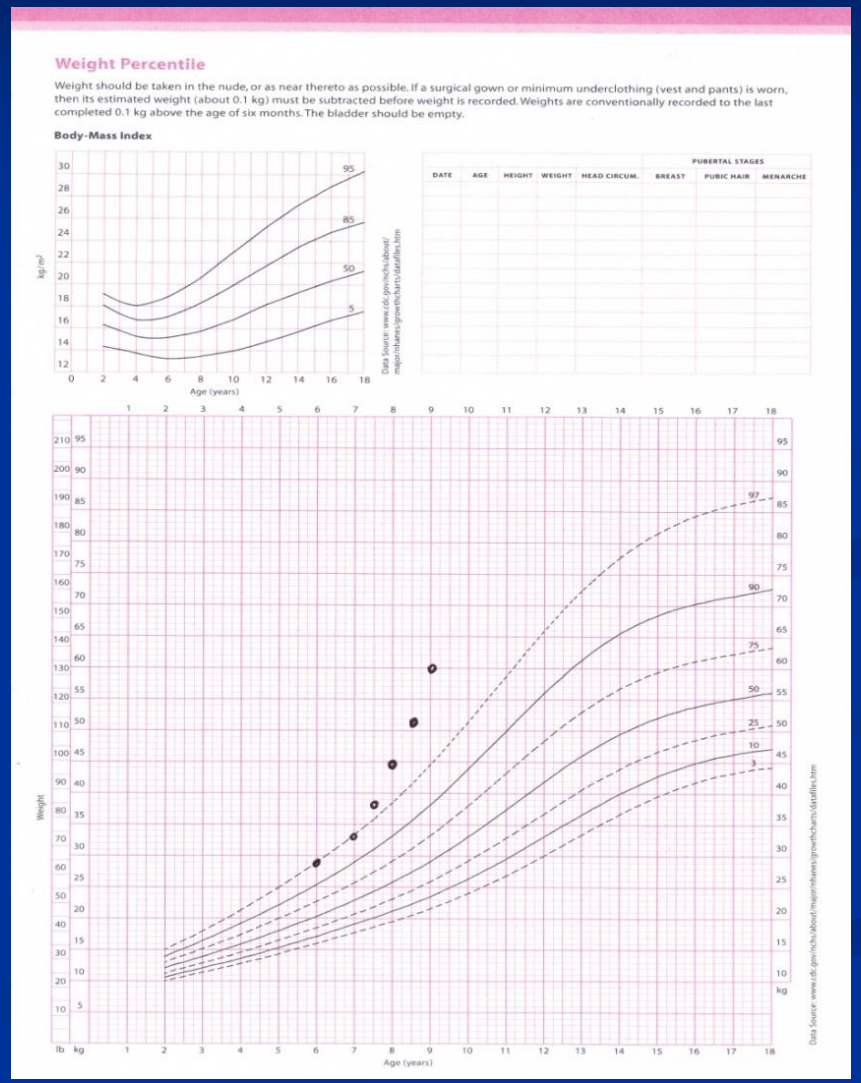


Basic Short stature work-up

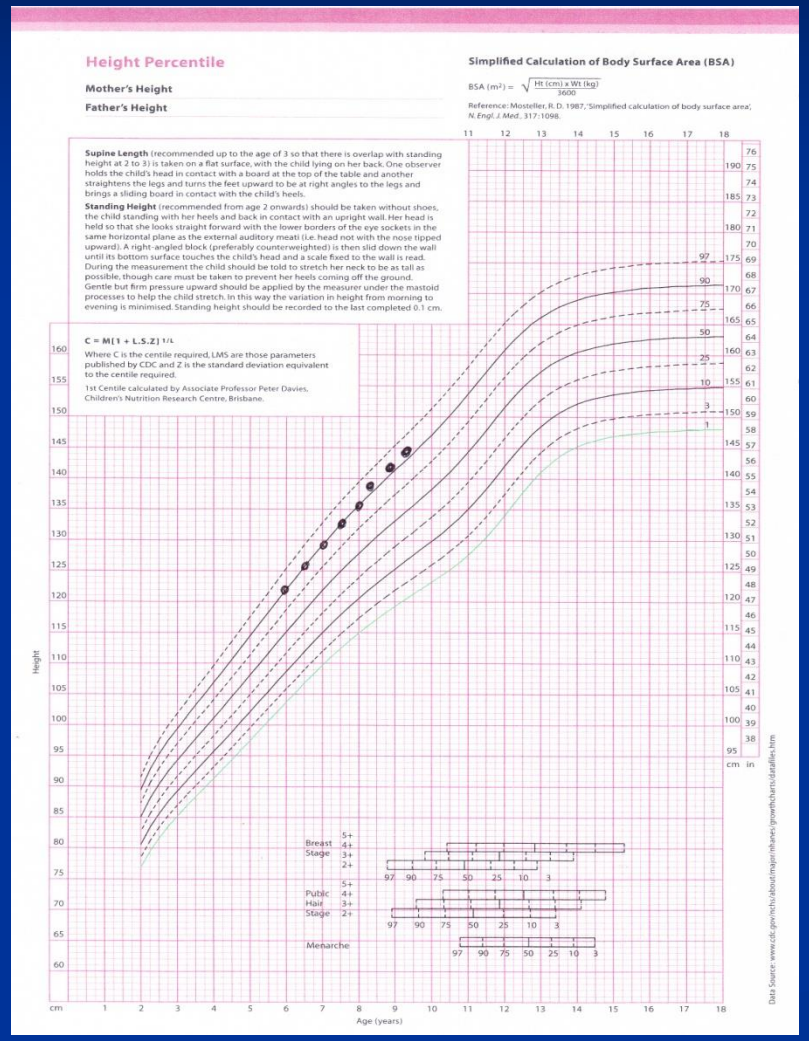
- Labs:
  - TFTs
  - Coeliac Screen
  - IGF-1
  - Electrolytes and renal
  - ESR
  - KARYOTYPE IN ALL GIRLS
- Radiology
  - Bone age

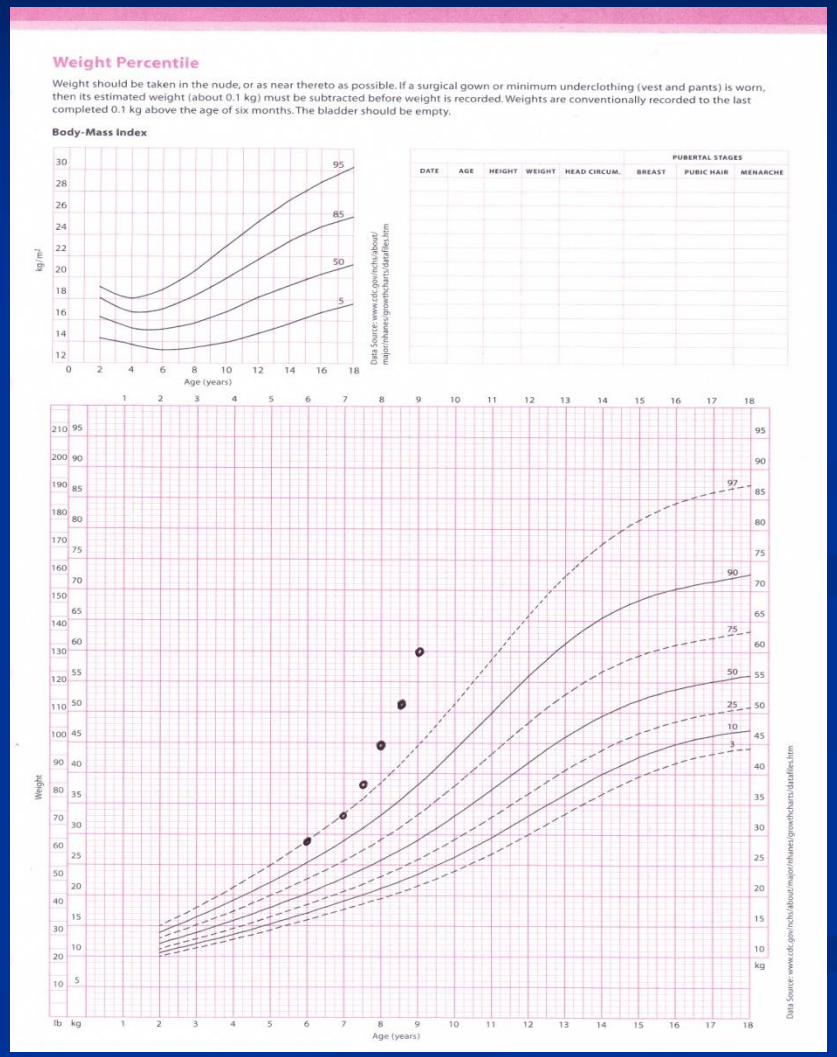
Cushing


Height Percentile
Mother's Height
Father's Height
Simplified Calculation of Body Surface Area (BSA)
Supine Length (recommended up to the age of 3 so that there is overlap with standing height at 2 to 3) is taken on a flat surface, with the child lying on her back. One observer holds the child's head in contact with a board at the top of the table and another straightens the legs and turns the feet upward to be at right angles to the legs and brings a sliding board in contact with the child's heels.
Standing Height (recommended from age 2 onwards) should be taken without shoes, the child standing with her heels and back in contact with an upright wall. Her head is held so that she looks straight forward with the lower borders of the eye sockets in the same horizontal plane as the external auditory meati (i.e. head not with the nose tipped upward). A right-angled block (preferably counterweighted) is then slid down the wall until its bottom surface touches the child's head and a scale fixed to the wall is read. During the measurement the child should be told to stretch her neck to be as tall as possible, though care must be taken to prevent her heels coming off the ground. Gentle but firm pressure upward should be applied by the measurer under the mastoid processes to help the child stretch. In this way the variation in height from morning to evening is minimised. Standing height should be recorded to the last completed 0.1 cm.
Where C is the centile required, LMS are those parameters published by CDC and Z is the standard deviation equivalent to the centile required.
1st Centile calculated by Associate Professor Peter Davies, Children's Nutrition Research Centre, Brisbane.
Height
Age (years)
Breast Stage
Pubic Hair Stage
Menarche


Weight Percentile
Weight should be taken in the nude, or as near thereto as possible. If a surgical gown or minimum underclothing (vest and pants) is worn, then its estimated weight (about 0.1 kg) must be subtracted before weight is recorded. Weights are conventionally recorded to the last completed 0.1 kg above the age of six months. The bladder should be empty.
Body-Mass Index
PUBERTAL STAGES
DATE
AGE
HEIGHT
WEIGHT
HEAD CIRCUM.
BREAST
PUBIC HAIR
MENARCHE
Age (years)
Weight

Obesity-Exogenous





PUBERTY

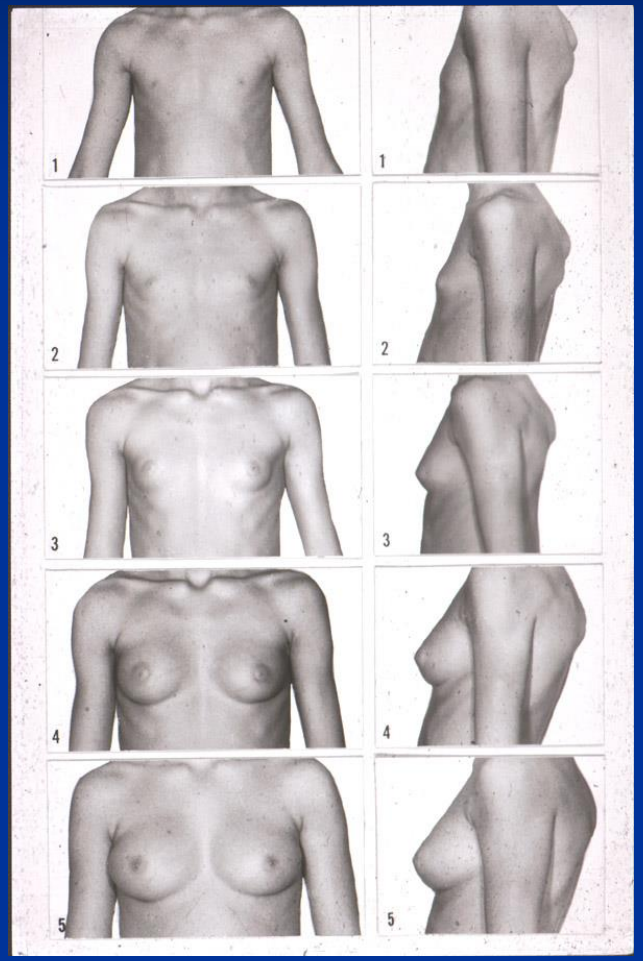
- Acquisition of reproductive capability
- Marked by:
  - Appearance of secondary sexual characteristics; (female breast development, sexual hair, male genital growth, male voice changes)
  - Growth spurt

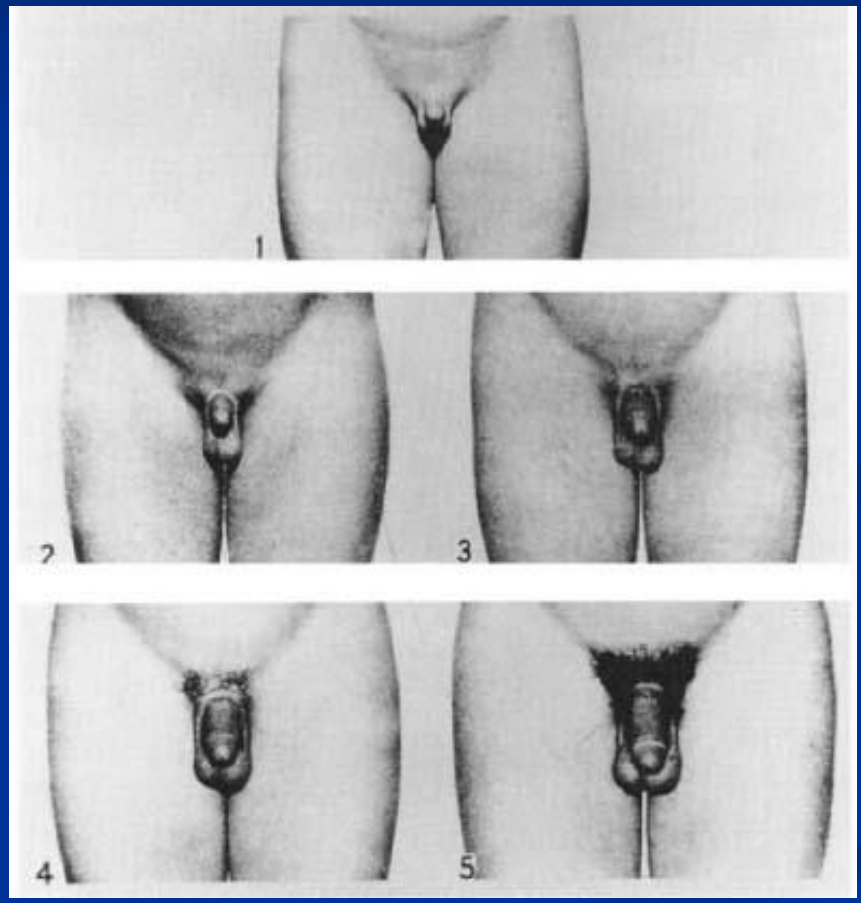
Puberty - physiology

- Increasing pulses of gonadotrophin releasing hormone from hypothalamus →
- ↑secretion of gonadotrophins - follicle stimulating hormone (FSH) and luteinizing hormone (LH) →
- ↑secretion of sex steroids – oestrogens and androgens → secondary sexual characteristics

Tanner Stages of puberty:

(I prepubertal-V fully pubertal)





Boys 2-18 years

Stages of Puberty

Ages of attainment of successive stages of pubertal sexual development are given in the Height Percentile chart.
The stage Pubic Hair 2+ represents the state of a child who shows the pubic hair appearance stage 2 but not stage 3 (see below).
The centiles for age at which this state is normally seen are given, the 97th centile being considered as the early limit, the 3rd centile as the late limit. The child's puberty stages may be plotted at successive ages (Tanner. 1962, *Growth at Adolescence*, 2nd edn.). Testis sizes are judged by comparison with the Prader orchidometer (Zachmann, Prader, Kind, Haffinger & Budliger. 1974, *Helv. Paed. Acta.* 29, 61-72).

Genital (Penis) Development

Stage 1. Pre-adolescent. Testes, scrotum and penis are of about the same size and proportion as in early childhood.
Stage 2. Enlargement of scrotum and testes. Skin of scrotum reddens and changes in texture. Little or no enlargement of penis at this stage.
Stage 3. Enlargement of the penis which occurs at first mainly in length. Further growth of the testes and scrotum.
Stage 4. Increased size of penis with growth in breadth and development of glans. Testes and scrotum larger; scrotal skin darkened.
Stage 5. Genitalia adult in size and shape.

Pubic Hair Development

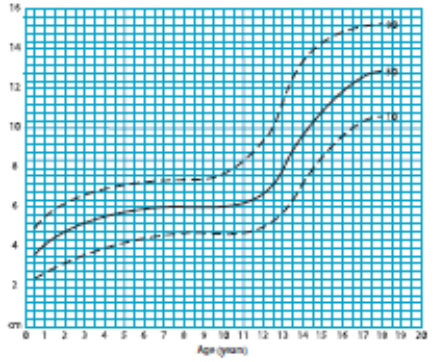
Stage 1. Pre-adolescent. The vellus over the pubes is not further developed than that over the abdominal wall, i.e. no pubic hair.
Stage 2. Sparse growth of long, slightly pigmented downy hair, straight or slightly curled at the base of the penis.
Stage 3. Considerably darker, coarser and more curled. The hair spreads sparsely over the junction of the pubes.
Stage 4. Hair now adult in type, but area covered is still considerably smaller than in the adult. No spread to the medial surface of thighs.
Stage 5. Adult in quantity and type with distribution of the horizontal (or classically 'feminine') pattern. Spread to medial surface of thighs but not up linea alba or elsewhere above the base of the inverse triangle (spread up linea alba occurs late and is rated stage 6).

Genital and Pubic Hair Development Stages

Stage 2 Stage 3 Stage 4 Stage 5

Stretched Penile Length

Measured from the pubo-penile skin junction to the tip of the glans (Shonfeld & Beebe. 1942, *Journal of Urology*, 48, 759-777).



Girls 2-18 years

Stages of Puberty

Ages of attainment of successive stages of pubertal sexual development are given in the Height Percentile chart.
The stage Pubic Hair 2+ represents the state of a child who shows the pubic hair appearance stage 2 but not stage 3 (see below).
The centiles for age at which this state is normally seen are given, the 97th centile being considered as the early limit, the 3rd centile as the late limit. The child's puberty stages may be plotted at successive ages (Tanner. 1962, *Growth at Adolescence*, 2nd edn).

Pubic Hair Development

Stage 1. Pre-adolescent. The vellus over the pubes is not further developed than that over the abdominal wall, i.e. no pubic hair.
Stage 2. Sparse growth of long, slightly pigmented downy hair, straight or slightly curled, chiefly along labia.
Stage 3. Considerably darker, coarser and more curled. The hair spreads sparsely over the junction of the pubes.
Stage 4. Hair now adult in type, but area covered is still considerably smaller than in the adult. No spread to the medial surface of thighs.
Stage 5. Adult in quantity and type with distribution of the horizontal (or classically 'feminine') pattern. Spread to medial surface of thighs but not up linea alba or elsewhere above the base of the inverse triangle (spread up linea alba occurs late and is rated stage 6).

Breast Development Stages

Stage 1. Prepubertal

Stage 2. Elevation of breasts and papilla

Stage 3. Further elevation and areola but no separation of contours

Stage 4. Areola and papilla form a secondary mound above level of the breast

Stage 5. Areola recesses to the general contour of the breast

Pubic Hair Stages

Stage 2

Stage 3

Stage 4

Stage 5

Puberty in boys

- Marked by testicular enlargement 4 cc or more
- Growth spurt at testicular size of 12 cc (late)

Normal Puberty in Girls

- First evidence usually **breast budding**
- Increase in growth velocity (**growth spurt** peak st3 breast development)
- Most growth occurs in first half of puberty
  - (prior to menarche), ave 4-6 cm afterwards
- Interval between pubertal onset and **menarche** about 2 years
- Av duration of pubertal development 3-4 yrs

Abnormal puberty

Boys

- Precocious if occurs

< 9 years

- Delayed if no puberty by 14 years

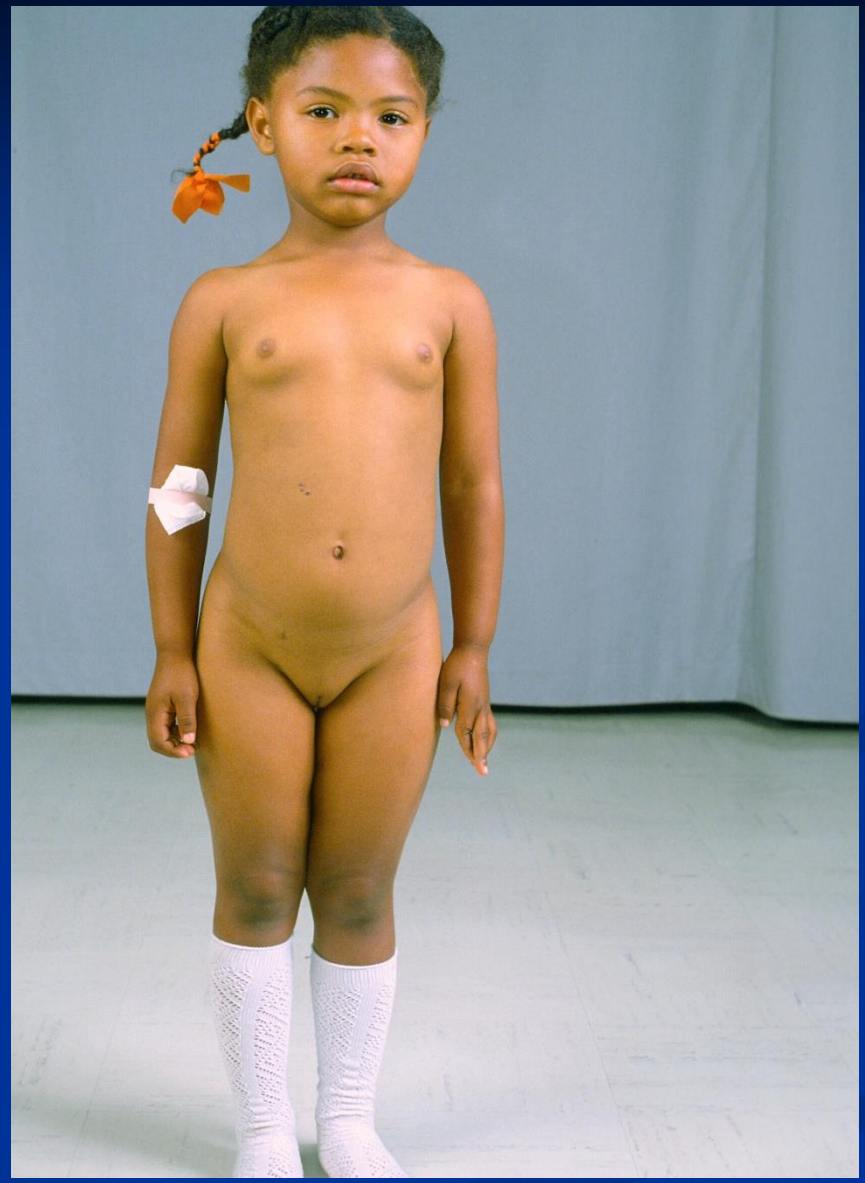
Girls

- Precocious if occurs

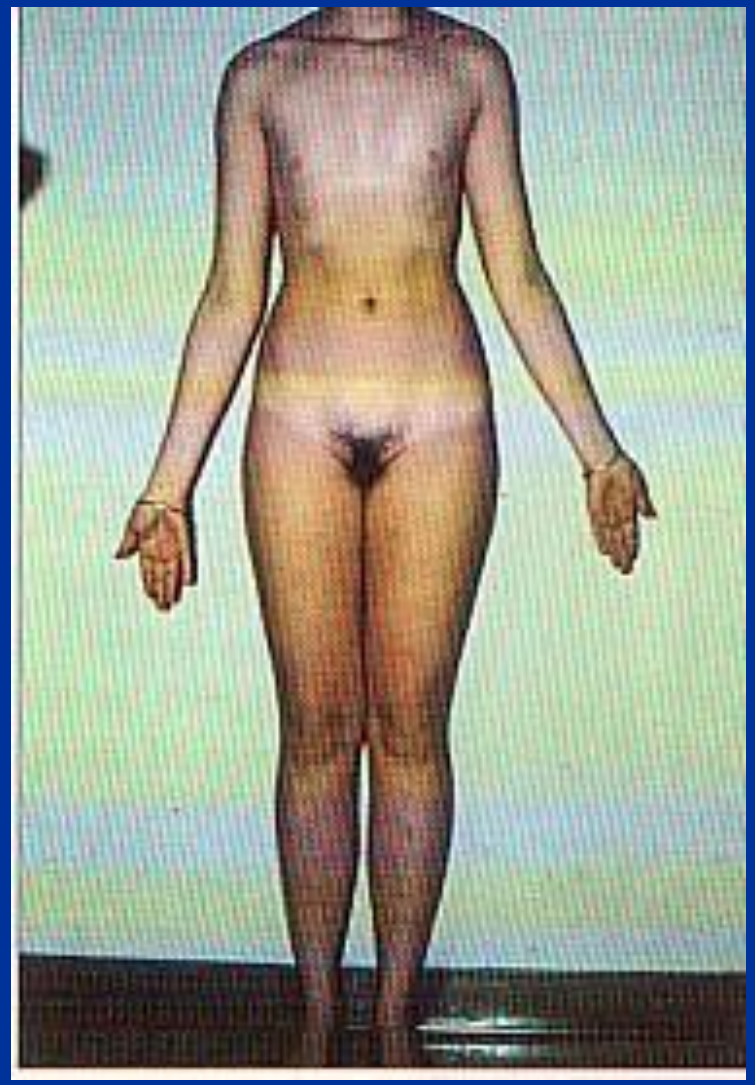
< 8 years

- Delayed if no breast budding by 13.5 years or no menses five years after thelarche

- 6 year old girl with breast growth for 6 months + growth spurt
- Investigations:
- FSH and LH in pubertal range
- Pelvic U/S: enlarged uterus with endometrial thickening
- Diagnosis: Central precocious puberty
- Brain MRI: hypothalamic hamartoma



Age 15



- Worried about lack of breasts and having not started her period
- Pubic hair present for 4 years
- Otherwise well
- Parents tall
- Ht 140 cm, prepubertal

- Investigations:
  - low oestrogen, high FSH and LH (hypergonadotrophic hypogonadism) ➔ end-organ failure
  - Karyotype: 45 XO
- Diagnosis: Turner Syndrome

- 50% of Turner karyotypes 45XO
  - Tend to have more phenotypic features
- 40% have no features other than delay in puberty +/- short stature

Common clinical issues related to growth and puberty

Premature adrenarche/pubarche

- production of adrenal androgens (adrenarche) causing appearance of pubic hair(pubarche) at < 8 yrs
- NOT indicative of true puberty if isolated (no other signs of puberty (increasing testes in boys, growth spurt, breast budding in girls)

Pubertal Gynaecomastia

- Breast growth in pubertal boys
- Common (40-70%)
- Usually minor and transient, may be asymmetrical
- May require plastic surgery if significant/permanent

Premature thelarche

- Isolated breast development, either unilateral/bilateral
- Relatively common in girls <2 yrs, characteristically waxing and waning in size
- Can occur later
- Not progressive and not accompanied by other signs of puberty

Thank You

Don't forget to take the session survey!