

Growth, Puberty and Hormones

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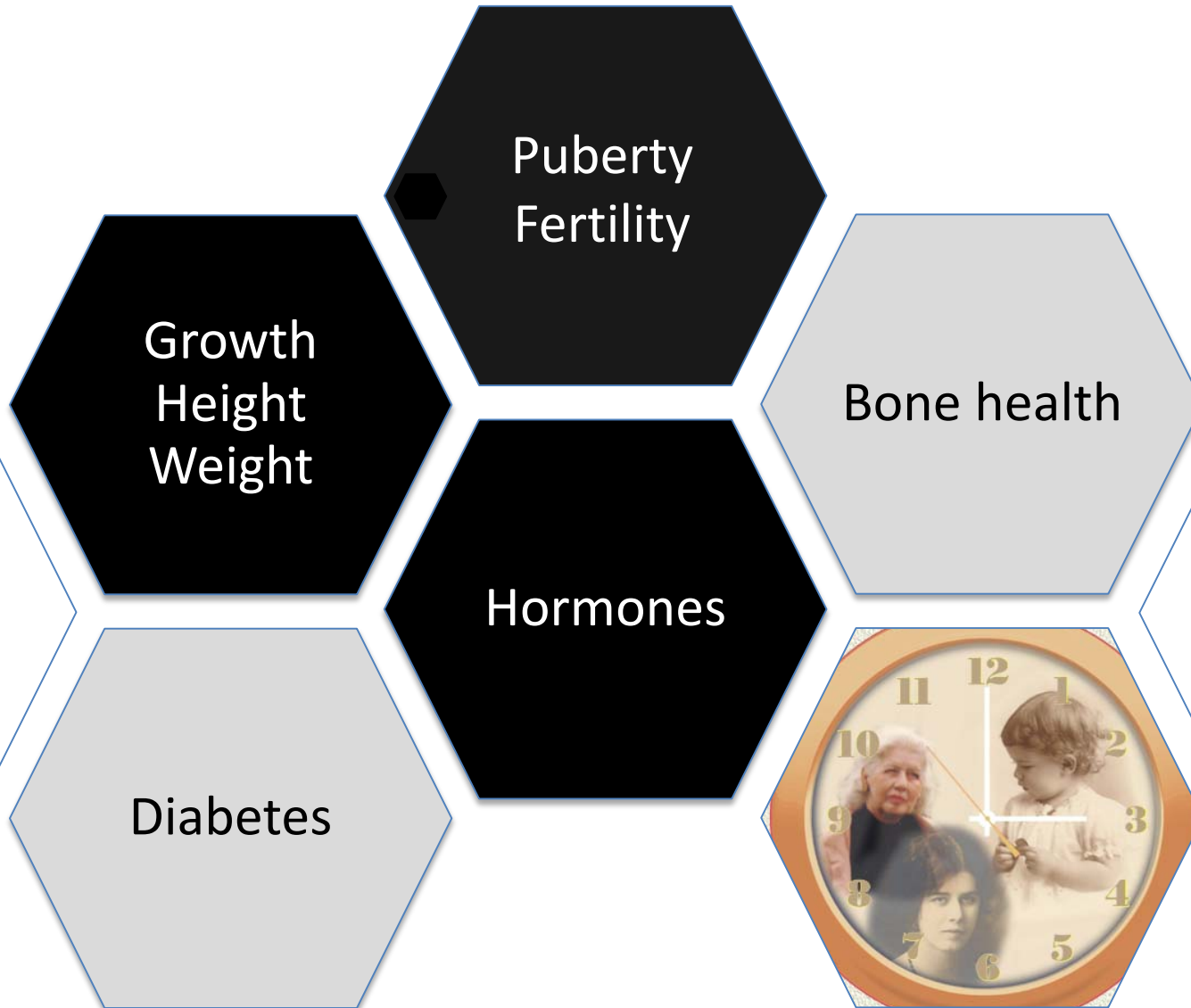
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Dr Claire Higham

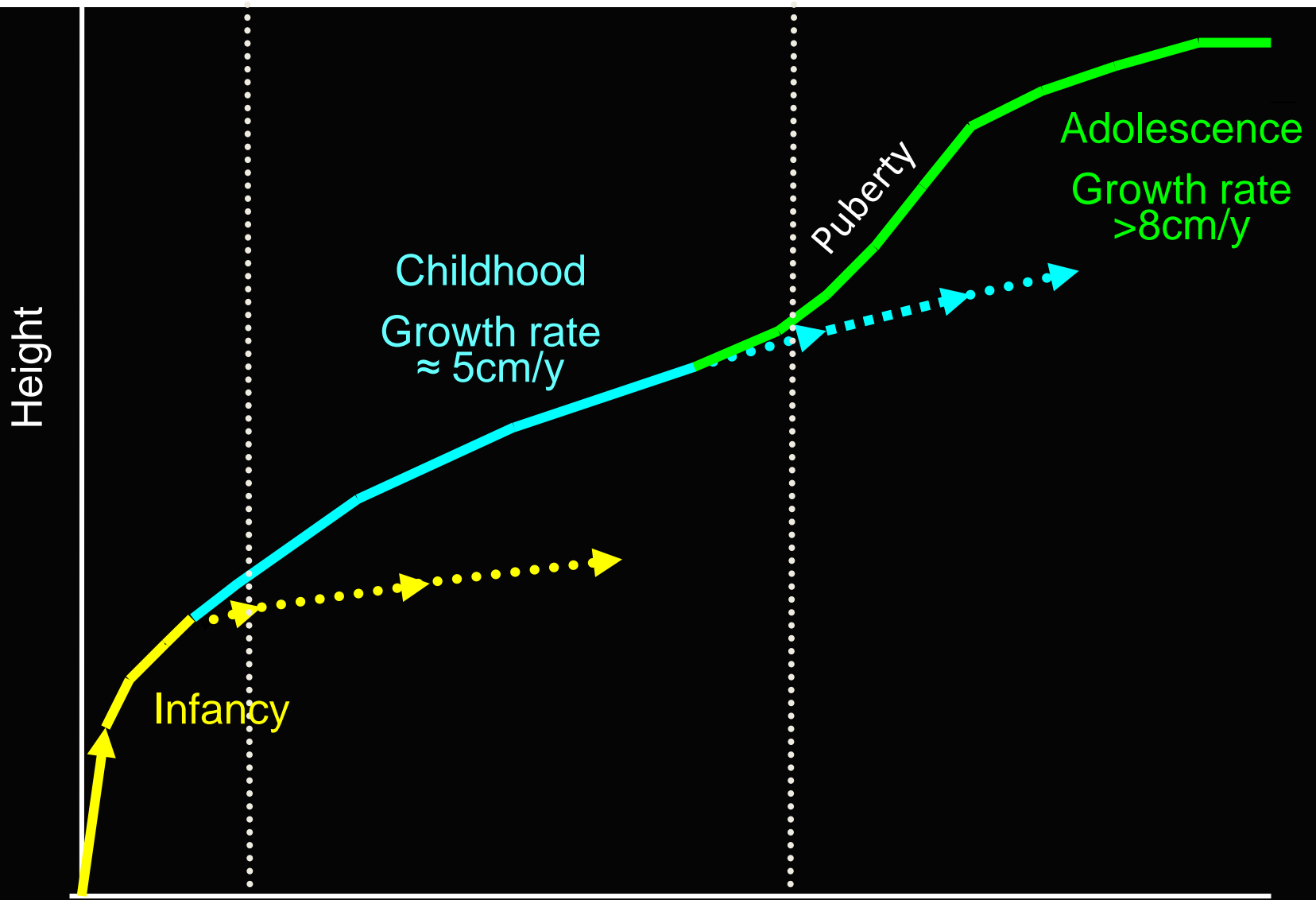
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This session



Growth from Birth to Adulthood



Birth	6m	1y	2y	6y	12y	20y
BW	> x2 BW	> x3 BW	x4 BW	> x6 BW	> x12 BW	x20 BW

Assessment of growth and puberty

weight

length/height

head size

puberty



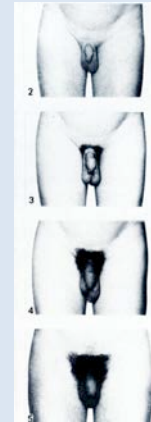
Accurate

Every appointment

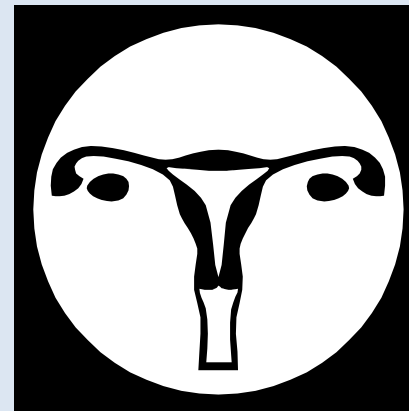
Plotted on growth charts

FA specific growth charts not available

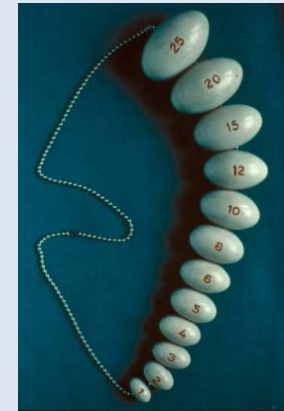
Secondary sex characteristics



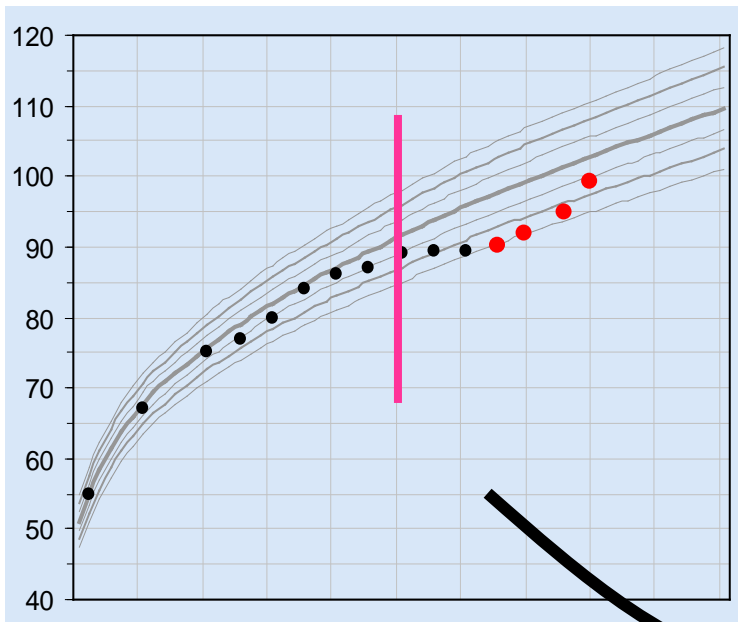
Ovaries



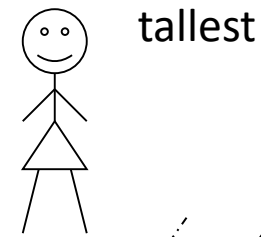
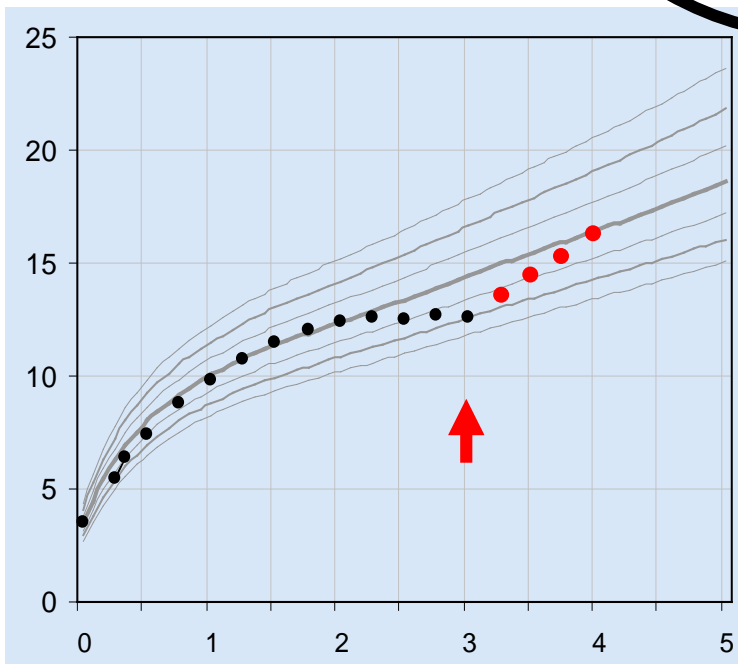
Testes



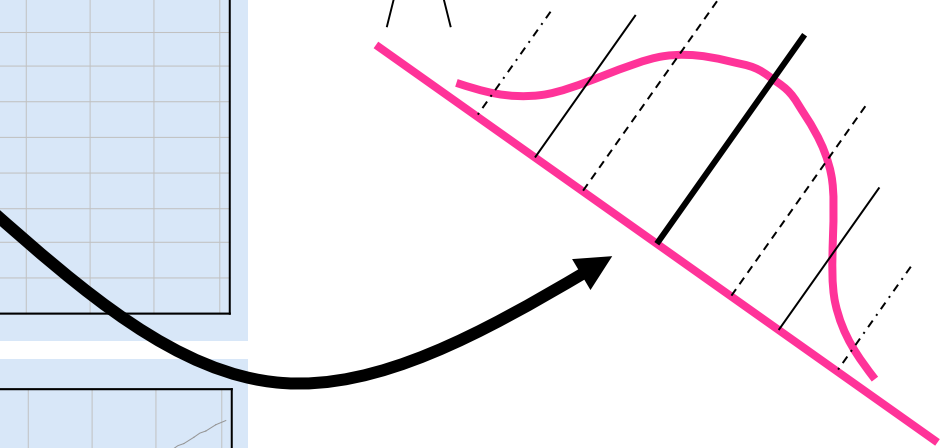
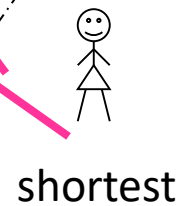
Height



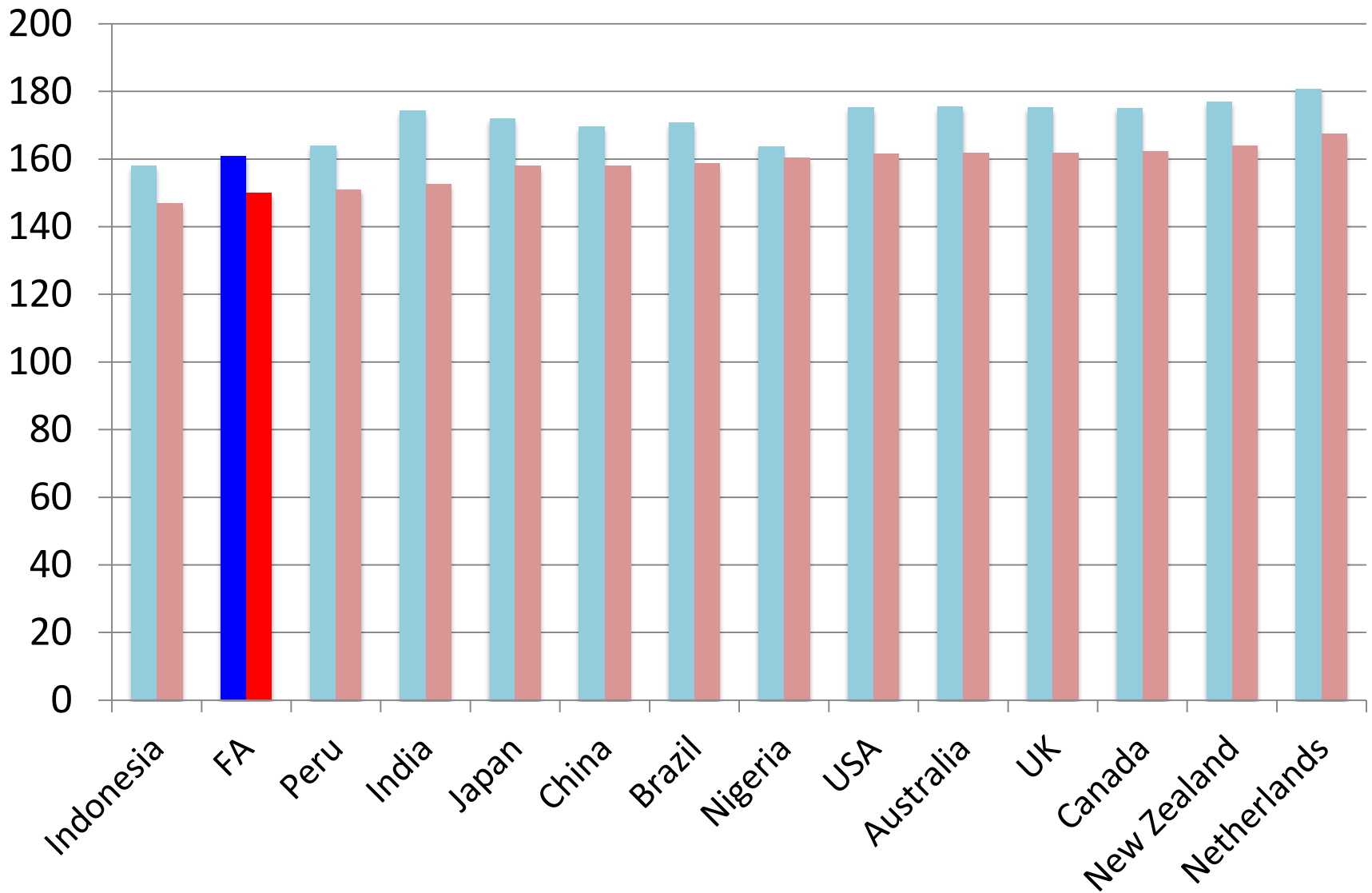
Weight



average



Height around the world and adults with FA



Women 150cm
Men 161cm



Being relatively short is an inherent feature of FA

Principal requirements for normal growth

1. Hormones

- Growth hormone
- Thyroxine
- Puberty hormones

2. Nutrition

Insufficient nutrition

- poor intake, e.g. anorexia
- malabsorption, e.g. coeliac disease

3. Intake must be in excess of consumption

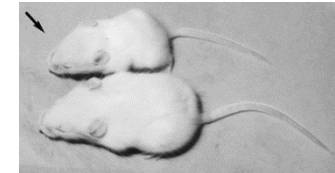
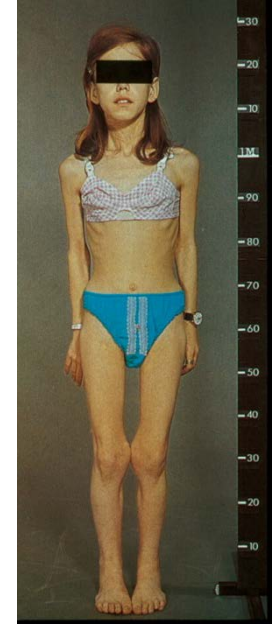
Excessive need for /over-consumption of nutrients

- chronic illness

4. Tissue synthesis must be in excess of degradation

Decreased tissue synthesis and excessive degradation

- steroid treatment
- diabetes mellitus



Individuals with FA are relatively short because ...

1. Hormones

- ✓ Growth hormone – but rare. Transplant is a potential risk factor
- ✓ Thyroxine – 37%
- ✓ Puberty hormones – 65%

2. Nutrition

Insufficient nutrition

- ✓ poor appetite, diarrhoea

3. Intake must be in excess of consumption

Excessive need for /over-consumption of nutrients

- ✓ chronic illness

4. Tissue synthesis must be in excess of degradation

Decreased tissue synthesis and excessive degradation

- ✓ steroid treatment – for graft vs host disease
- ✓ diabetes mellitus – rare

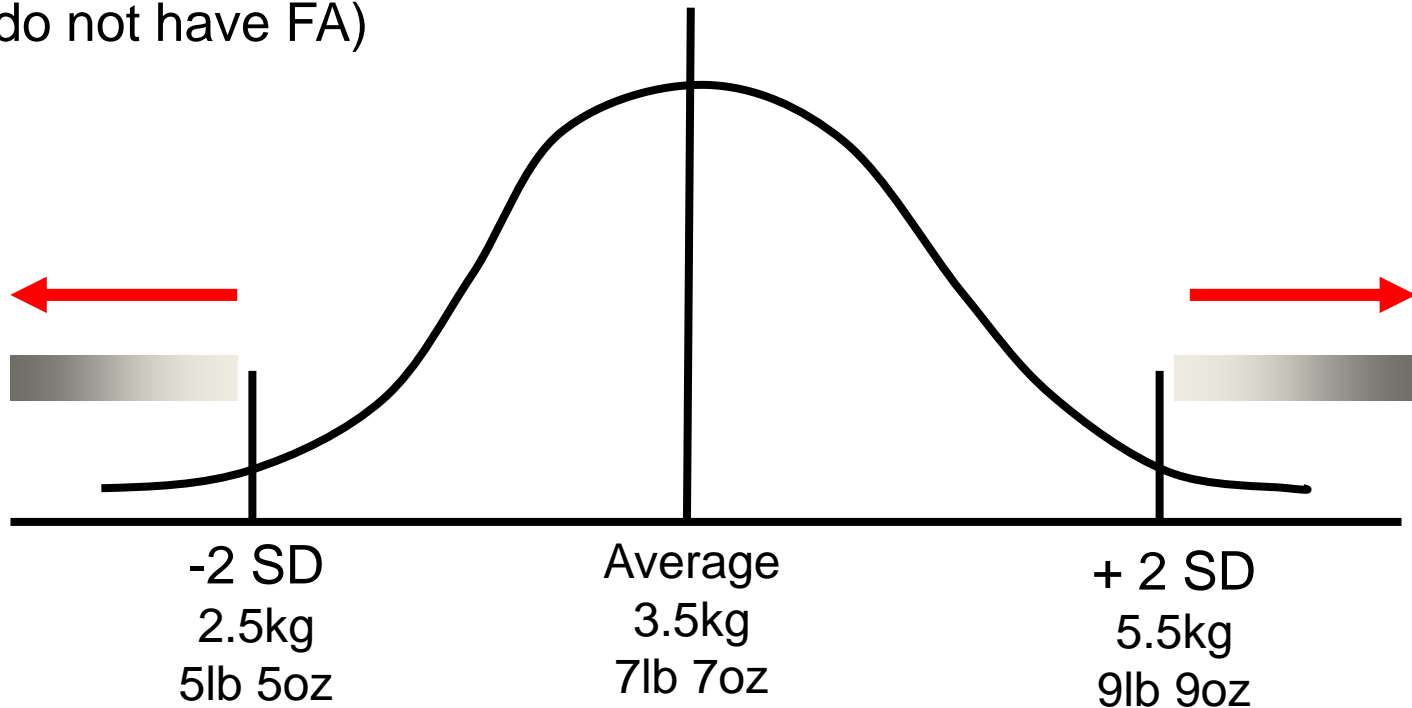
5. Born small – part of the condition and related to gene defects

Birth weight

50% of children with FA are small at birth



25% will catch-up after birth
(vs 90% of children who do not have FA)



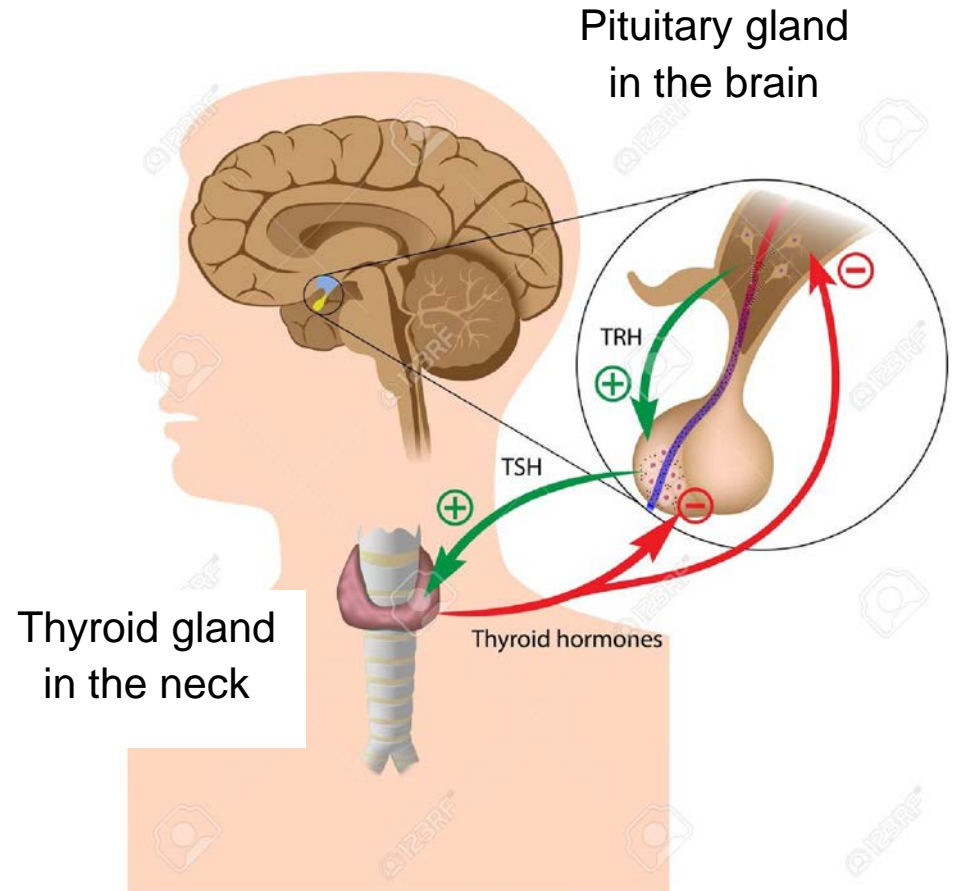
Hormone deficiencies in people with FA

1. Growth hormone from pituitary

- Rare
- Poor growth in height

2. Thyroid hormone – thyroxine

- Upto $\frac{1}{3}$ of individuals
- Cells in thyroid gland lost possibly from unrepaired DNA damage (oxidative injury)
- Pituitary gland affected due to radiotherapy
- Poor growth in height
- “Slow”, sleepy, sluggish



Hormone deficiencies in people with FA

1. Growth hormone from pituitary gland
2. Thyroid hormone – thyroxine

1. Puberty hormones

Puberty can happen

- Too early – ‘precocious’
Brain signals prematurely

- Too late

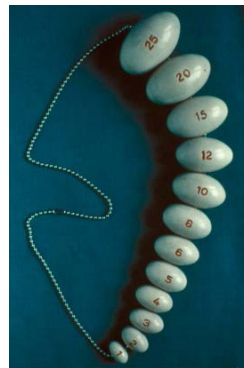
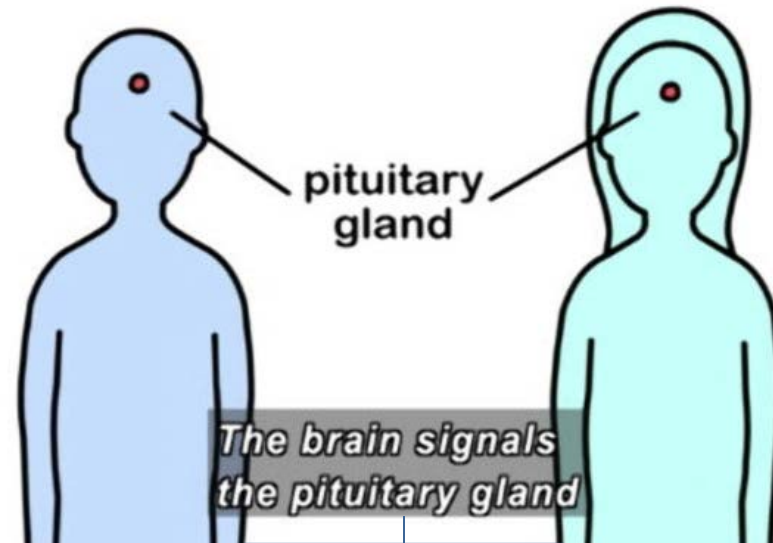
Pituitary delayed

or

Testes or ovaries
don't work well



Fertility can be affected

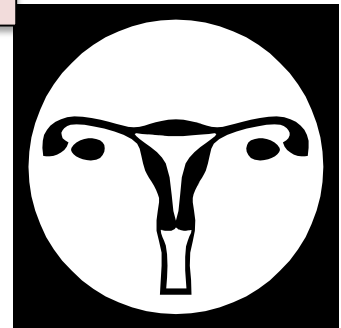


Testes develop

Produce testosterone

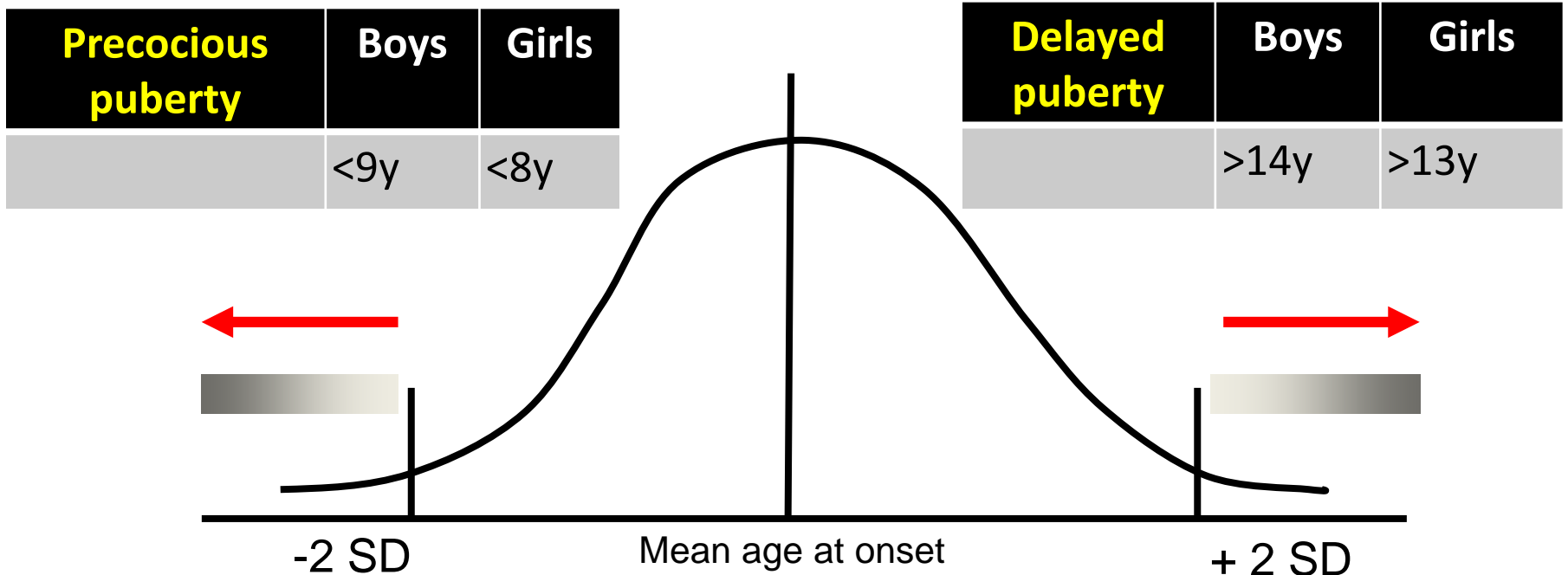
Ovaries develop

Produce oestrogen



Variations in the timing of onset of puberty

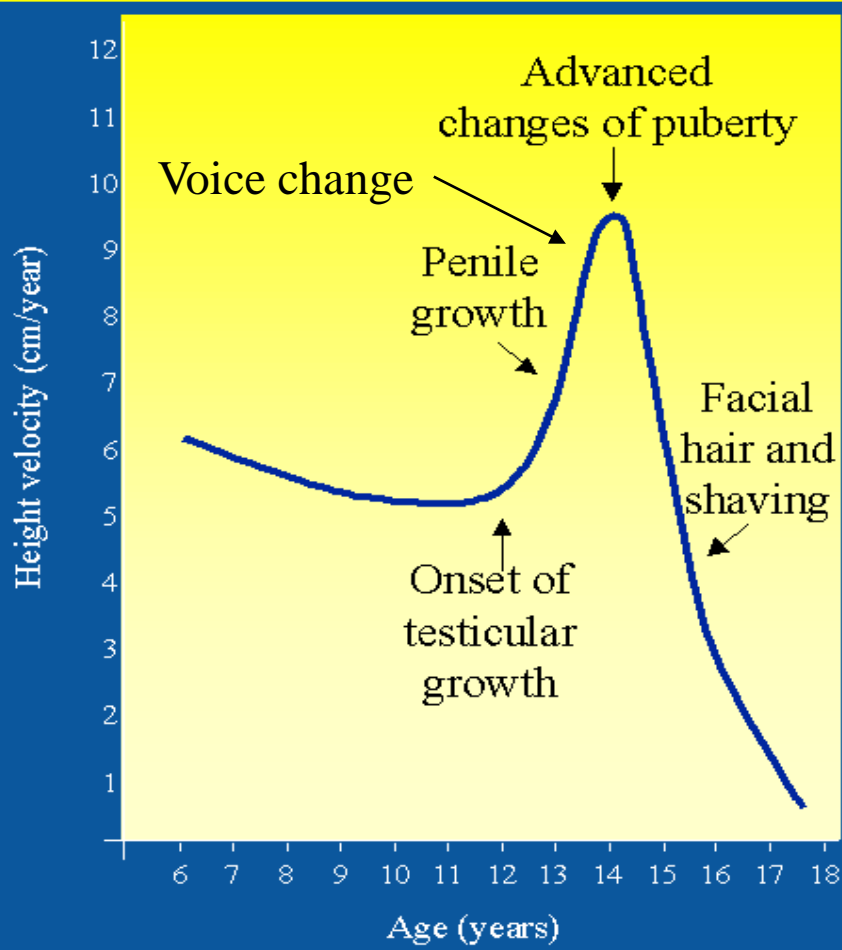
Normal	Boys	Girls
Age of onset of puberty: Average Range	12y 9.5 – 13.5y	11y 8.5 – 13y



The sequence of changes during puberty and in relation to the pubertal growth spurt

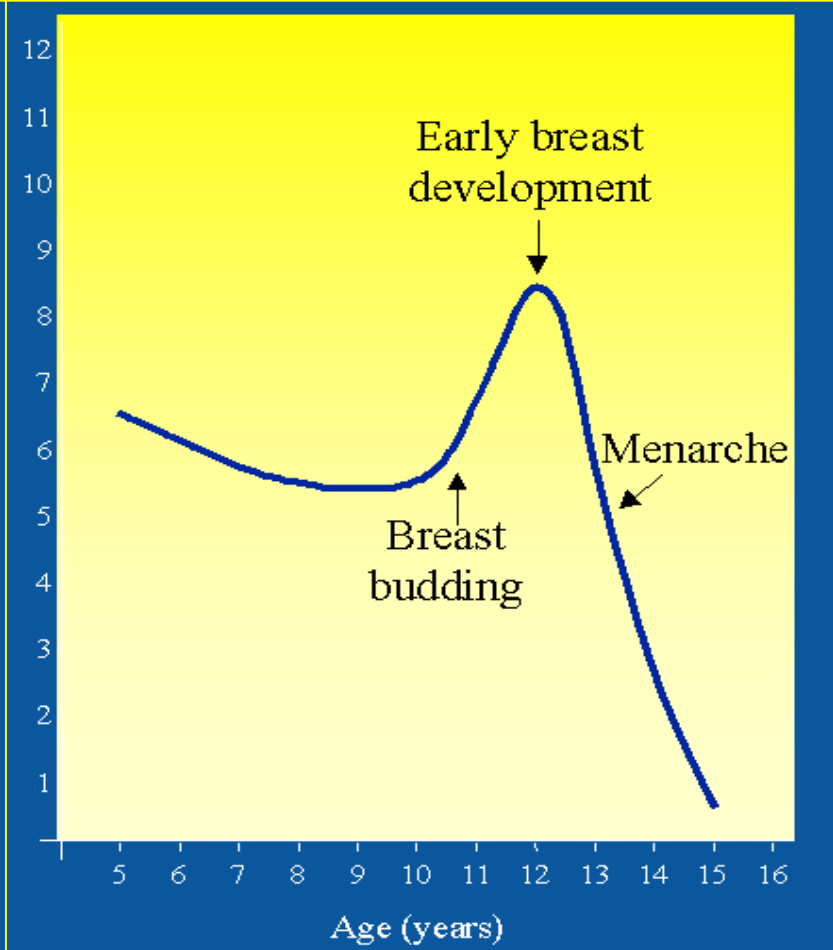
Boys:

Fastest growth at G3-4, testes 10-12ml;
mean age 13.5y

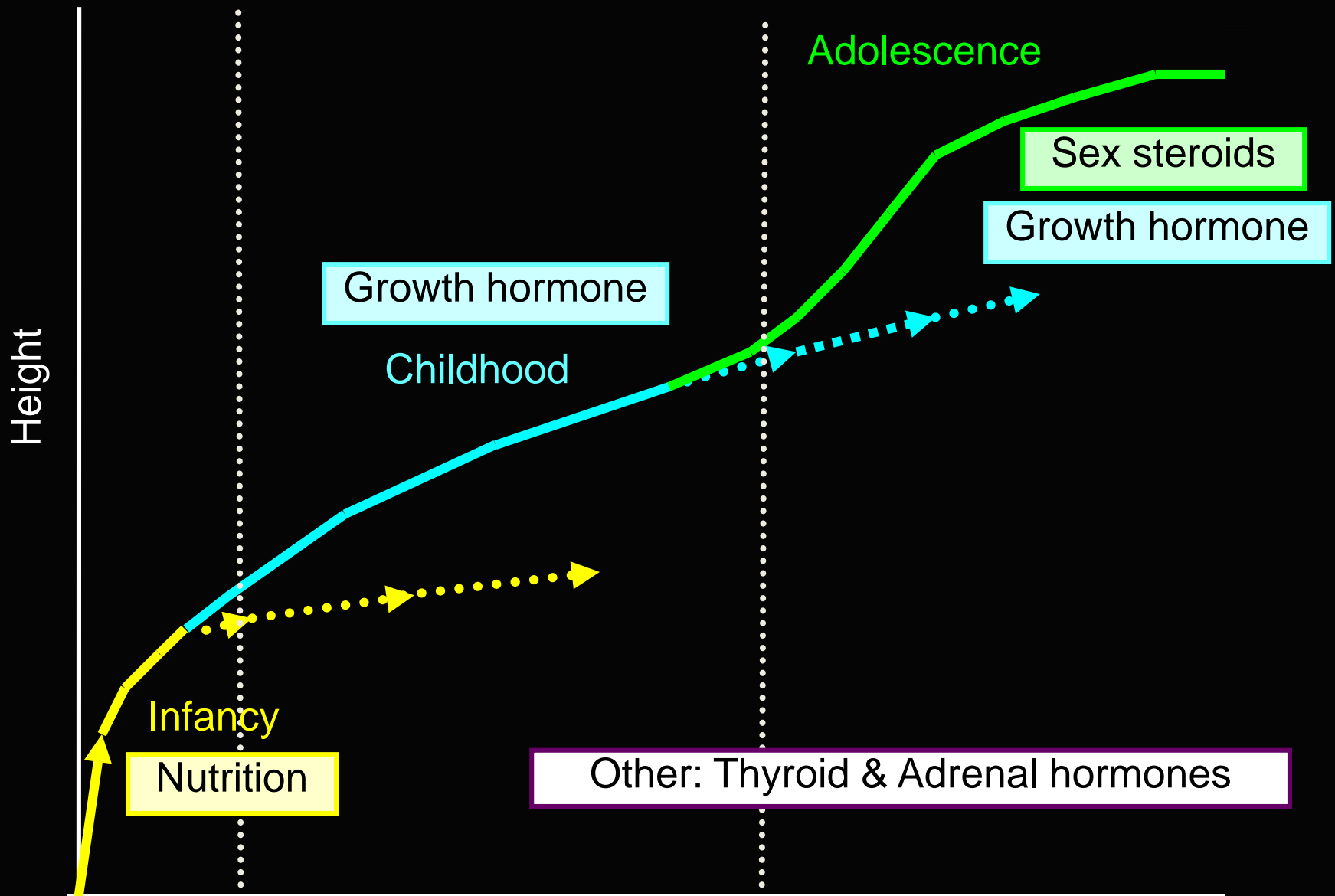


Girls:

Fastest growth at B2-3;
mean age 11.5y



Key factors required for growth after birth



Any questions so far ?

