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THE SHORT OF IT

A Practical Approach to Short Stature

Paul Hofman
2009

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Why bother?

3% of people will always be below the third percentile!
In the vast majority of cases there is no medical problem...

And yet

People complain of being short even when normal e.g. on the 25th percentile

There is great anxiety among many families with short children...especially where the parents are also short.

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- Is being vertically challenged a handicap?
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SHORT STATURE and Society

Tall stature is valued – perhaps even more in shorter societies!

Males are taller than women in general and historically in most societies hold power.

Ruling classes possibly due to better nutrition and genetic selection are taller.

Thus height has become associated with power, intelligence and dominance.

In the USA height has been named as near top of the list in traits people value when choosing a mate

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Heightism is as more prevalent than racism!

Short people are often the butt of jokes (look online at some of the websites!)

Many positive comments associated with tall stature
look 'up' to
stand 'tall'
holding in 'high' regard

Whereas there are negative associations with small size
talking 'down'
possessing 'short-comings'
'short man syndrome'

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Heightism

There is no denying that we place a high premium on height, be it social, sexual, or economic, and our preference for height pervades almost every aspect of our lives.

Heightism is "one of the most blatant and forgiven prejudices in our society." Economist John Kenneth Galbraith (height 6'8").

1996 essay titled 'My Inner Shrimp' by Gary Trudeau creator of *Doonesbury* – "for the rest of my days I will be a recovering short person". His final height was over 184cm but was very short as a child.

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The long term cost of short stature

For some short stature has a profound effect – even when they are not substantially small. Some have published books on their experiences.

Two very worthwhile books to read

Beyond Measure: A Memoir About Short Stature and Inner Growth by Ellen Frankel

Size matters: How Height Affects the Health, Happiness and Success of Boys - and the Men They Become by Stephen S Hall.

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Short stature – long term sequelae

Despite the clear prejudice against small stature are there negative consequences for the individual?

Much of the justification for height promoting therapies is based on the studies demonstrating long term negative psychological outcomes for short children.

However these studies are often biased and focus on selected subjects.

Many recent studies have demonstrated no clear psychological changes solely due to reduced height.

Empirical status of stature-related stereotypes

STEREOTYPE	EVIDENCE
Children and adults with SS are more poorly adjusted psychologically	Generally supported by analogue based research. Not supported by general population or clinic based studies
Children and adults with SS are treated poorly due to their stature	Mixed results from analogue studies. Evidence of teasing and juvenilization from clinic based studies.
Short men are less attractive and desirable to women as dates or husbands	Generally supported by analogue research. Limited support in population studies: effect attenuated when controlling for confounding variables.

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Empirical status of stature-related stereotypes

STEREOTYPE	EVIDENCE
Children and adults with SS do less well at school/are less intelligent	Generally supported by analogue studies. Not supported by general population or clinic based studies of children or adults.
Adults with SS hold lower status occupations and are paid less.	Supported by analogue studies. Limited support in population based studies: effect attenuated when controlling for confounding variables.

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Pathological Short Stature

Short stature or even more sensitive, poor growth velocity can reflect underlying pathology.

Linear growth is extremely sensitive to any alteration in either the external or internal environment.

Poor growth is therefore a very sensitive, objective parameter indicating an underlying problem but it is not very specific.

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Short Stature Definitions

Short Stature is defined arbitrarily as a height less than the 3rd PC

Dwarfism is defined as height less than 3 S.D. below the mean (0.5 PC)

Short Stature Definitions

Relevance

Growth Hormone Therapy in NZ is available for children with

- GH deficiency
- Turner Syndrome (Growth velocity <25th PC)
- Renal Failure (Growth velocity <25th PC, Height <3rd PC)
- Prader Willi Syndrome (Growth velocity <25th PC)
- **Severe Short Stature (Height <-3 SD and growth velocity <25th PC)**

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Short Stature vs Failure to Thrive

Failure to thrive is defined as failure to gain adequate weight which can lead to a secondary failure in height growth.

Important to identify as the aetiologies are very different to short stature.

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SHORT STATURE

Common perceived problem in the community and most common referral to paediatric endocrinologists.

Boys referred >> girls

95+% of short stature is NORMAL VARIANT

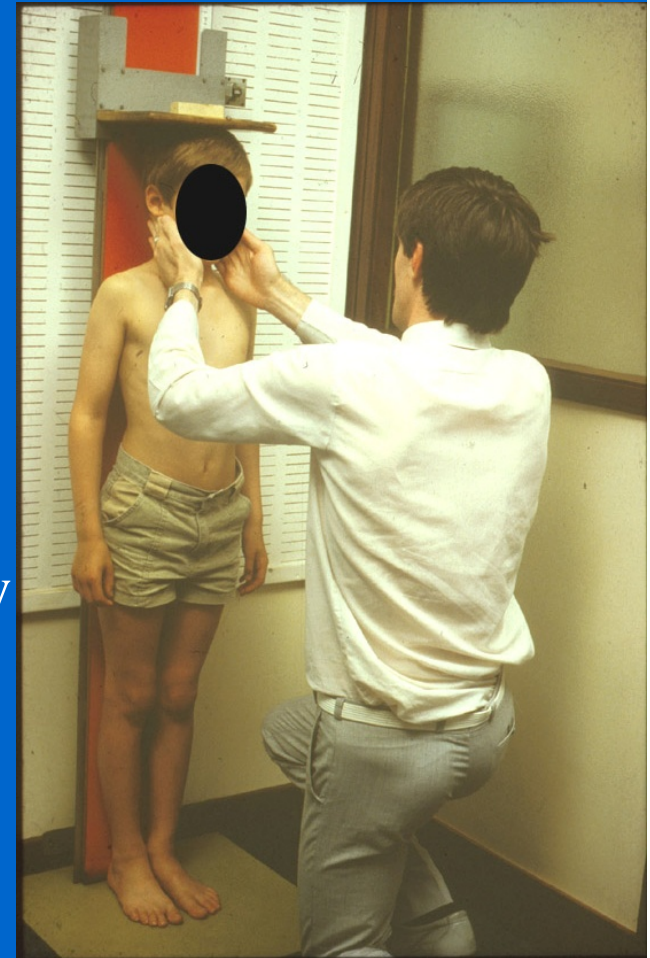
IT IS ESSENTIAL FOR DOCTORS DEALING WITH CHILDREN TO UNDERSTAND NORMAL VARIANT GROWTH AND ARE DISTINGUISH THIS FROM PATHOLOGICAL CAUSES.

SHORT STATURE Awareness!

To identify short stature you have to

1) MEASURE the patient

- have an accurate stadiometer
 - firm base
 - rigid measuring bar
 - standard length rod for checking height consistency
- know how to measure reliably and consistently
 - no shoes
 - straight legs
 - no tip toeing/ slouching
 - face in neutral plane with slight traction



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SHORT STATURE Awareness!

2) PLOT the data!

No one can accurately assess short stature by eyeballing a child.

Appropriate growth charts are needed. These available free on the APEG website (Australasian Paediatric Endocrine Group) under clinical resources.

<http://www.apeg.org.au/>

Looking at growth patterns is more useful than one point ie measure and plot all children at least annually if possible.

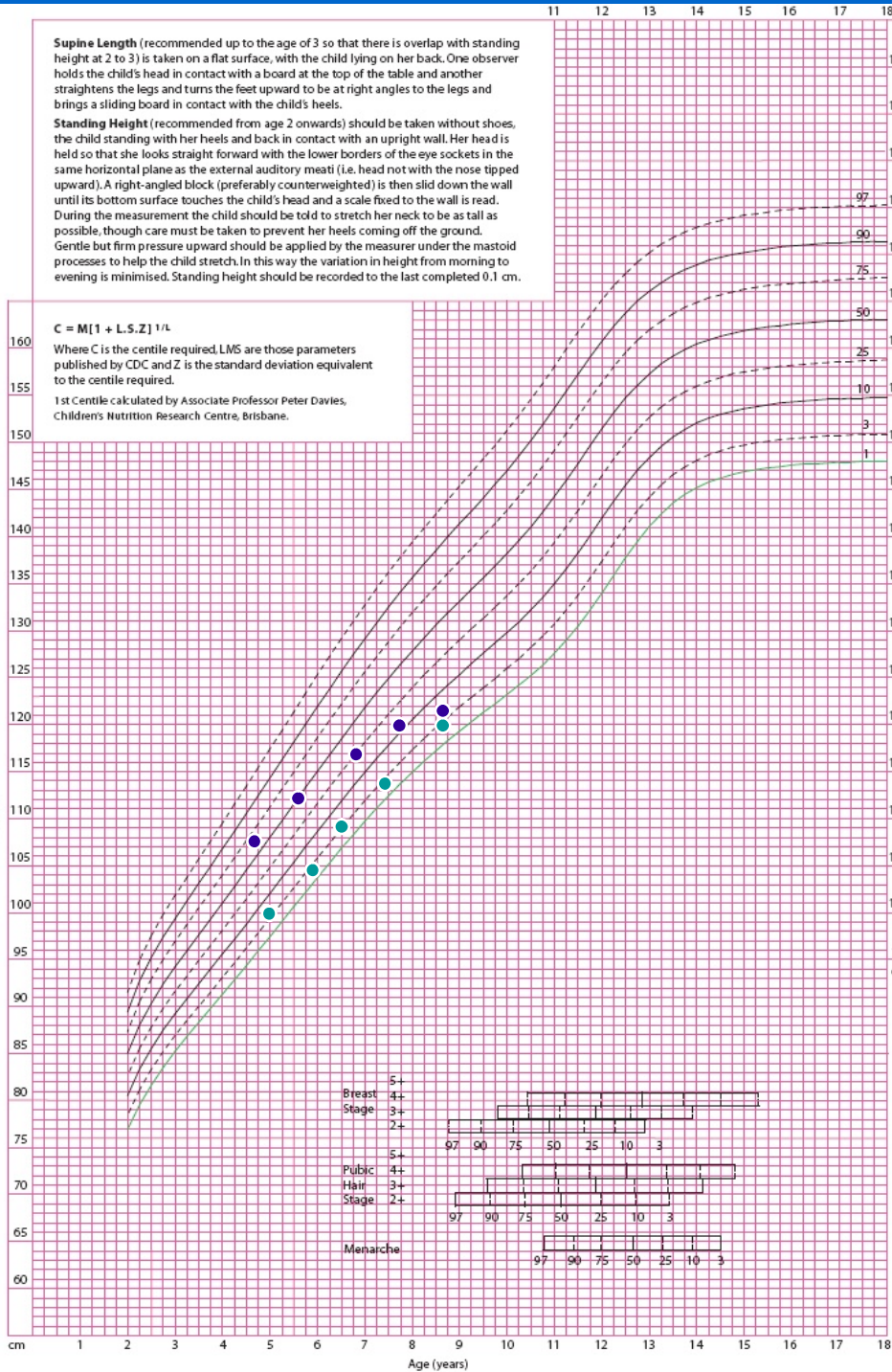
Supine Length (recommended up to the age of 3 so that there is overlap with standing height at 2 to 3) is taken on a flat surface, with the child lying on her back. One observer holds the child's head in contact with a board at the top of the table and another straightens the legs and turns the feet 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.

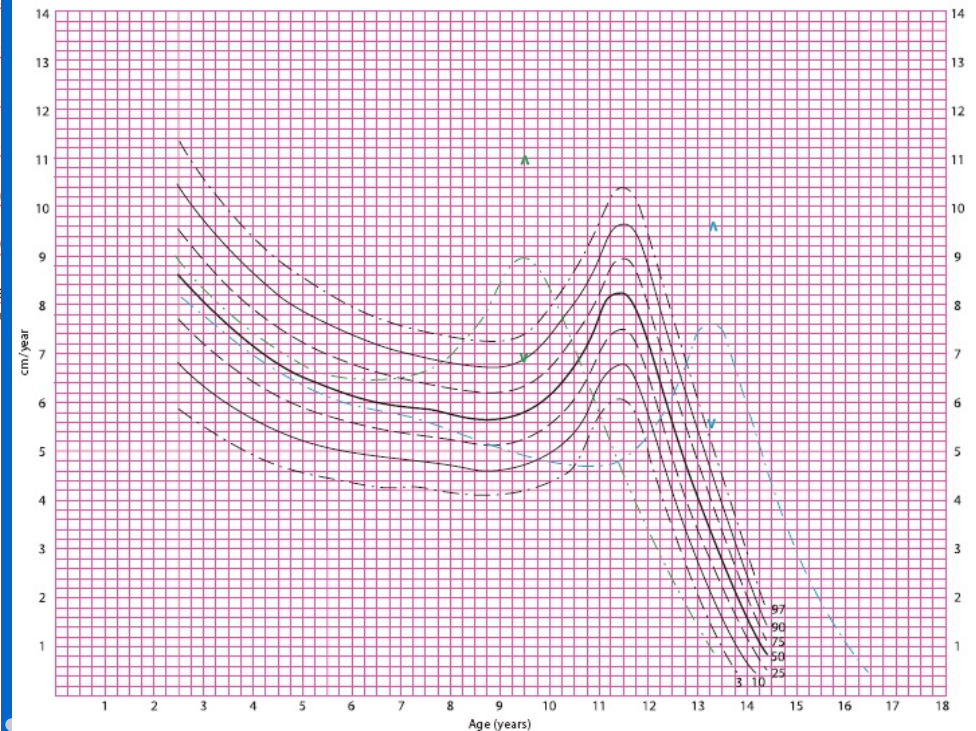
$$C = M[1 + L.S.Z]^{1/4}$$

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.



Girls 2-18 year Growth Charts



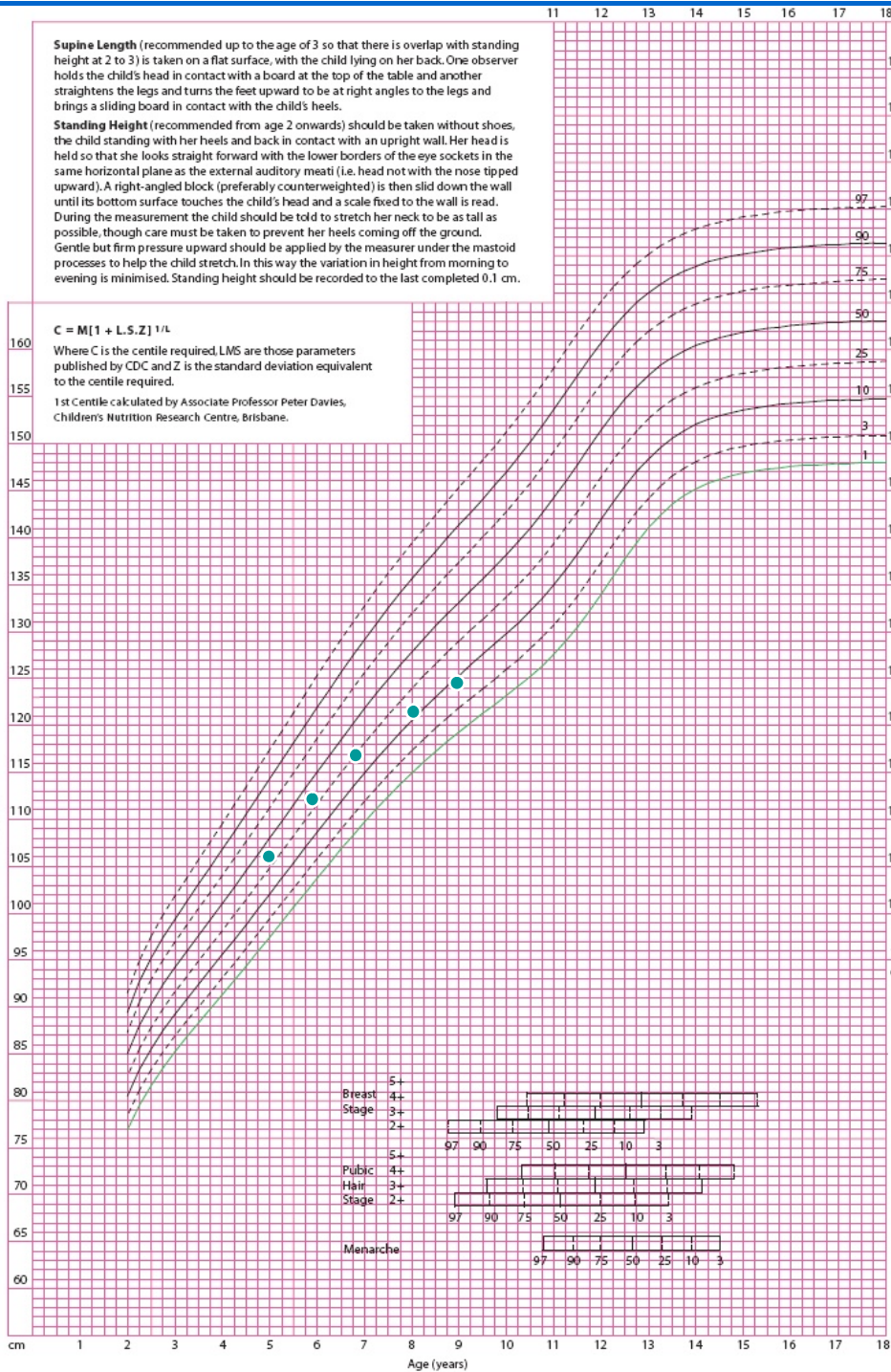
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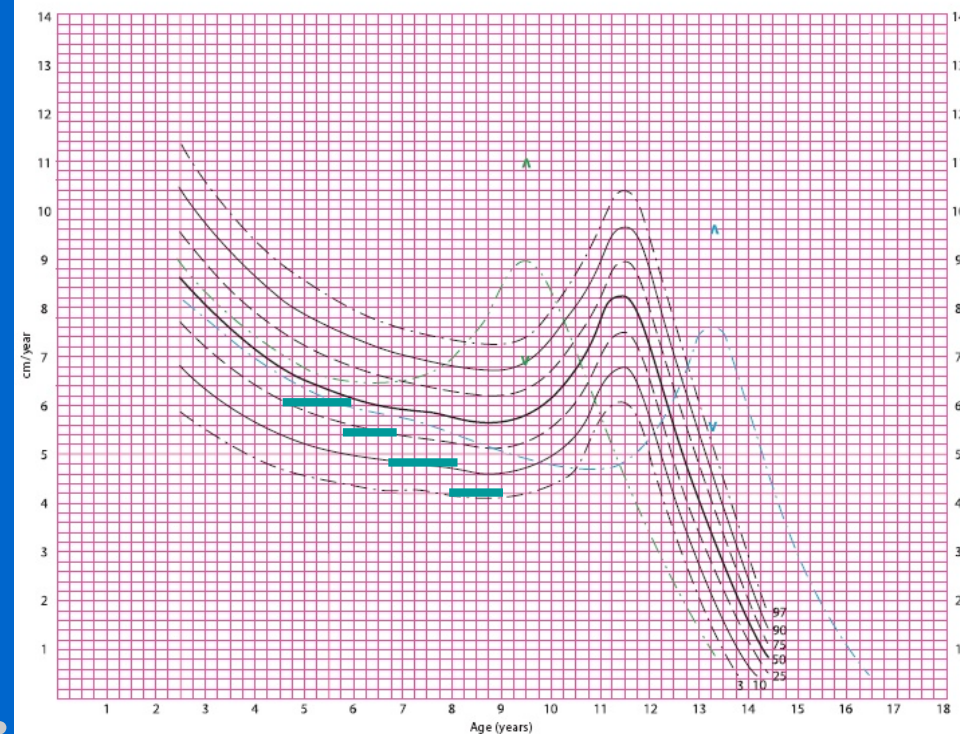
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Girls 2-18 year Growth Charts



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Normal Growth

REQUIREMENTS for growth/ anabolism

- Nurturing, caring environment
- Adequate nutritional supply and the ability to digest and absorb the food (ie the child should be well nourished)
- Appropriate hormonal milieu
- Appropriate extracellular and intracellular environment.

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Normal Variant Growth- Constitutional Delay of Growth and Development

- Child's height is well below that expected from the mid parental height.
- **Growth velocity is normal** (ie >25th %ile)
- Normal birth weight
- Family hx of pubertal delay and late growth common.
- **Bone age delayed** (although usually not more than 2 years)
- Final adult height normal (usually close to mid parental height)

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Normal Variant Growth- Constitutional Delay of Growth and Development

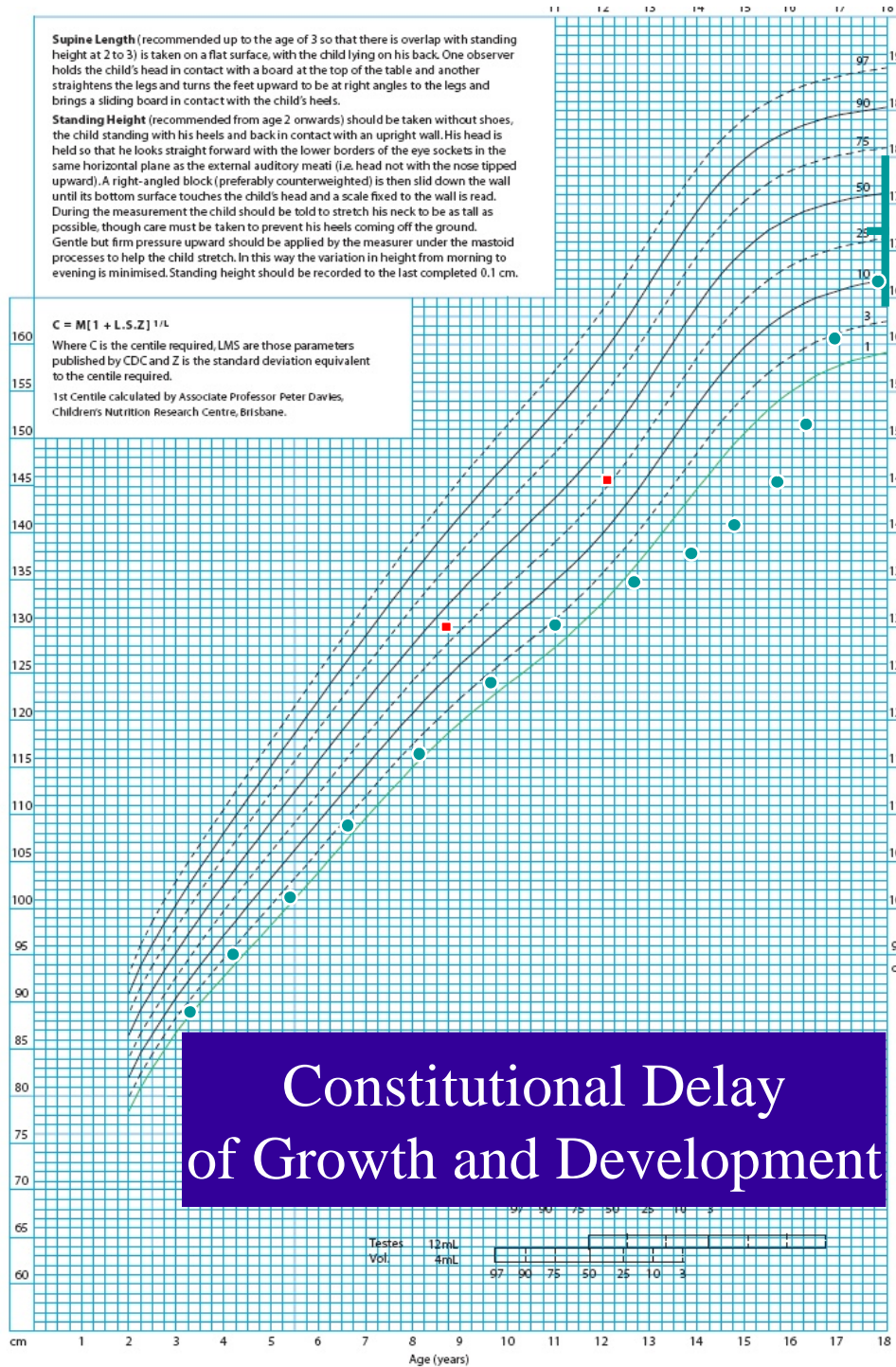
Characteristic history is normal growth for the first 6-9 months after which they cross percentiles downwards for 6-12 months. They then grow parallel to this percentile until late childhood.

Puberty is entered late with a corresponding delay in epiphyseal fusion and consequently extra growth.

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Constitutional Delay of Growth and Development

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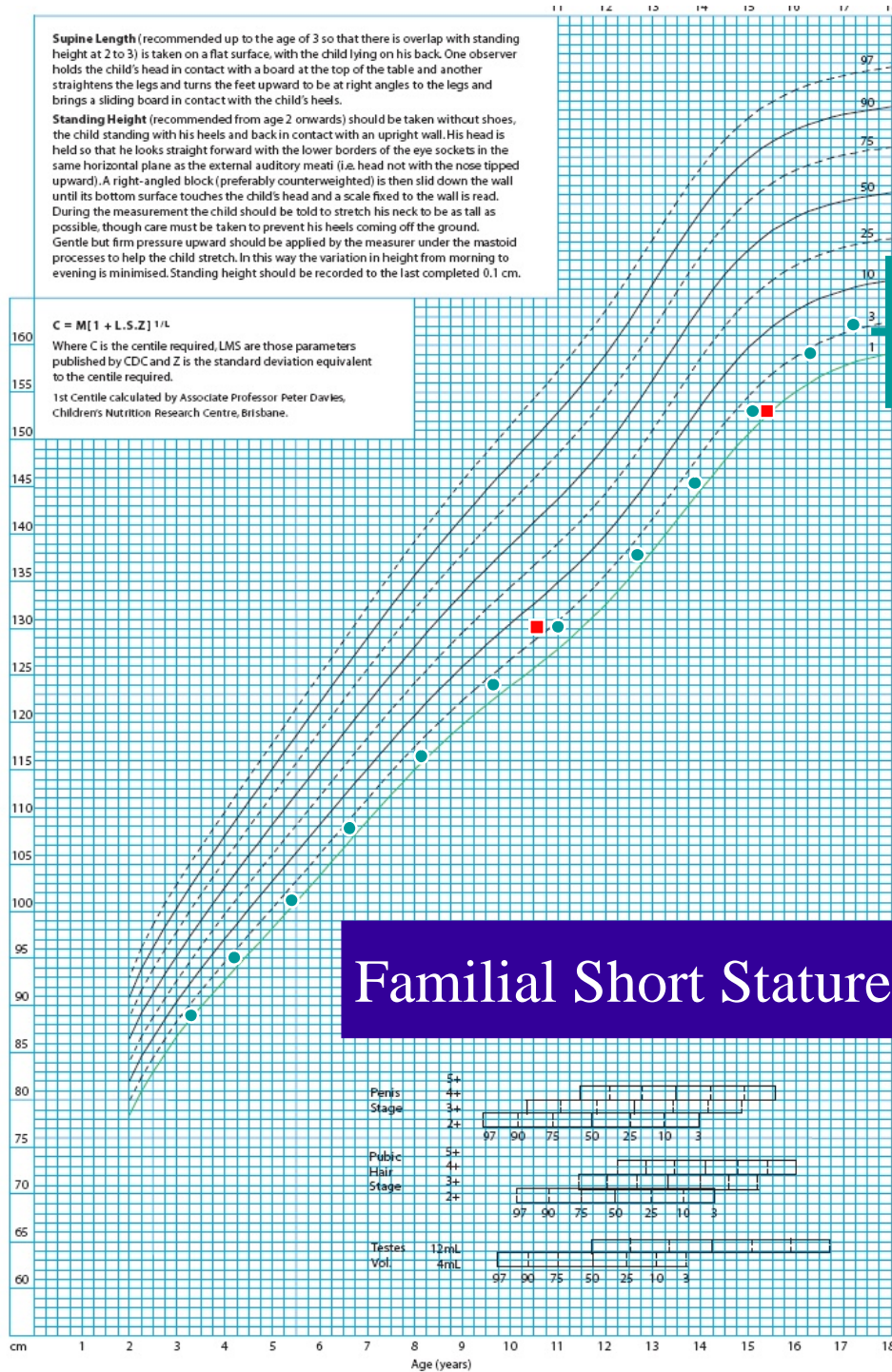
Normal Variant Growth- Familial Short Stature

- Short parents (beware one short parent - can represent a dominant genetic problem).
- Normal growth velocity (ie >25th PC)
- Normal birth weight
- Bone age appropriate for chronological age.
- Final height consistent with mid parental height.
- Typically these children grow down to their height percentile over the first 6 months after birth and then grow along and parallel to this percentile.

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Short Stature - Classification

Large number of causes

All pathological causes of short stature will cause a poor growth velocity (<25th PC) and a crossing of percentiles downwards on a growth chart.

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Short Stature - Classification

Proportionate (ie leg length to body length and arm span are normal)

Disproportionate (shortening of either limbs or trunk)

Almost always due to a skeletal dysplasia (eg hypo and achondroplasia)

A large no. of conditions, which combined are still uncommon.

Define by measuring armspan (= height \pm 5cm) or sitting height (easier to measure and charts readily available)

Short Stature - Proportionate

Post Natal

Normal Variant

Pathological

- Nutritional (starvation commonest cause of SS worldwide)
- Metabolic esp metabolic acidosis
- Chronic Illness ie renal, cardiac, GI, respiratory, neurological.
- Severe neglect can cause psychosocial dwarfism
- Chronic Drug Intake mainly glucocorticoid tx.

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Short Stature - Proportionate

Prenatal

Intrauterine Growth Retardation

uteroplacental insufficiency

congenital infection

syndromal

Chromosomal (Turner syndrome (XO) commonest cause of pathological SS in girls (1:2000 female births - always consider in girls!))

Antenatal Drug abuse (esp. alcohol)

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Short Stature - History

Growth pattern

Antenatal history and **birth weight**

Hx of any chronic illness/ head trauma or medication

Hx of malnutrition/ neglect

Family Hx

Mid-parental height $\frac{\text{Mother's ht} + \text{Father's ht} \pm 13\text{cm}}{2}$

95% confidence intervals are $\pm 8\text{cm}$

Hx of delayed puberty

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Short Stature - Examination

Measure and plot height (plus other previous measurements if available)

Limb asymmetry/disproportion

Pubertal Status

Dysmorphism

Neurocutaneous stigmata

Evidence of organ system disease

Short Stature - Investigation

Standard Investigation

BONE AGE (skeletal survey occ. required if a skeletal dysplasia is suspected)

If there is an abnormal growth pattern (measurements at least 4 months apart (preferably 6-12 months apart).

Thyroid function tests

FBC + ESR

U+Es, Creatinine, (LFTs)

Urinalysis/ pH

Karyotype (girls)

Coeliac screen

(faecal spec/ LFTs/ capillary blood gas)

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Short Stature - Investigation

If growth failure persists refer to a paediatrician/
endocrinologist

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Short Stature - Diagnosis

A pathological growth pattern can be established from growth data, if necessary taken over a 4-12 month period.

Plotting previous growth data will often help establish the length of time the problem has been present.

A normal growth velocity **over a 12 month period** virtually excludes a pathological cause for short stature.

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Treatment Options

- 1) Remove underlying cause for the growth failure if identified (eg inhaled steroids)
- 2) Growth hormone (only proven growth promoter). Specialist only and restricted use by PHARMAC.
- 3) Tamoxifen/ antioestrogen (in peripubertal boys). Blocks epiphyseal fusion. Early data looks promising but no final height data. Should be specialist only.
- 4) Testosterone – inducing puberty in severely constitutionally delayed boys. Doesn't increase height but gets them there more quickly. Specialist only.

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Summary

- 1) Measure and plot children at least annually
- 2) A normal growth velocity essentially excludes significant pathology.
- 3) Always calculate the midparental height and ask about a history of pubertal timing.
- 4) A bone age is the only 'must do' investigation. Waiting until you can calculate a growth velocity before doing other tests is otherwise appropriate.
- 5) In short girls growing poorly ALWAYS do a karyotype.