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Klotz Communications 2017: From the shortest to the tallest

How to investigate a child with excessive growth?

Que faire devant un enfant trop grand ?

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Abstract

The diagnostic approach to tall stature in children is based on collecting birth data (macrosomia), sizes and family puberty, a family history of constitutional or pathological tall stature, search for a delay of development, dysmorphia, disproportion, analysis of the growth velocity (normal or accelerated), general examination and assessment of puberty, and bone age. When there is a history of psychomotor retardation, a family history of pathological tall stature, or a disproportion in the clinical examination, the genetic causes of tall stature will be mentioned. The most frequent causes are Marfan syndrome and similar, Sotos syndrome, Beckwith-Wiedemann syndrome, Klinefelter syndrome, and MEN2B. There are many genetic syndromes with tall stature, justifying consultation with the geneticist. When the speed of growth is accelerated, first of all it evokes puberty and early pseudopuberty, obesity and acromegaly. Finally, when the growth velocity is regular, and the parents are of tall stature, it evokes constitutional tall stature: this is the most frequent diagnosis, to retain after having rejected pathological tall statures.

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Keywords: Tall stature; Growth velocity; Marfan syndrome; Sotos syndrome; MEN2B

Résumé

La démarche diagnostique devant une grande taille chez l'enfant s'appuie sur le recueil des coordonnées de naissance (macrosomie ?), des tailles et pubertés familiales, des antécédents familiaux de grande taille constitutionnelle ou pathologique, la recherche d'un retard de développement, d'une dysmorphie, d'une disproportion, sur l'analyse de la vitesse de croissance (normale ou accélérée), sur l'examen général et pubertaire, et sur la réalisation d'un âge osseux. Lorsqu'il existe des ACTD de retard psychomoteur, une histoire familiale de grande taille pathologique, ou une disproportion à l'examen clinique, les causes génétiques de grande taille seront évoquées. Les causes les plus fréquentes sont le syndrome de Marfan et apparentés, le syndrome de Sotos, le syndrome de Wiedemann-Beckwith, le syndrome de Klinefelter, la MEN2B. Il existe de nombreux syndromes génétiques avec grande taille, justifiant d'une consultation avec le généticien clinicien. Lorsque la vitesse de croissance est accélérée, on évoque en premier lieu les pubertés et pseudopubertés précoces, l'obésité, l'acromégalie. Enfin, lorsque la vitesse de croissance est régulière, et les parents de grande taille, on évoque la grande taille constitutionnelle : il s'agit du diagnostic le plus fréquent, à retenir après avoir éliminé les grandes tailles pathologiques.

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Mots clés : Grande taille ; Vitesse de croissance ; Syndrome de Marfan ; Syndrome de Sotos ; MEN2B

1. Definitions and initial management

1.1. Definitions

Tall stature is defined as a size ranging beyond the threshold of +2 standard deviations of the average population size for the age

and gender. In France, a size greater than +2 standard deviations at adult age corresponds to more than 187 cm for a man and more than 175 cm for a woman. In the Netherlands, the same statistical thresholds lead to a size greater than 194.5 cm and 180 cm, and in Korea to 180 cm and 166.5 cm respectively (Fig. 1) [1].

A height beyond +2 standard deviations from the genetic target height (the average of the parental heights SD score) (Fig. 1), or an acceleration of growth (arbitrarily a statural height gain of more than 0.5 to 1 standard deviation) also merit medical assessment (Fig. 2).

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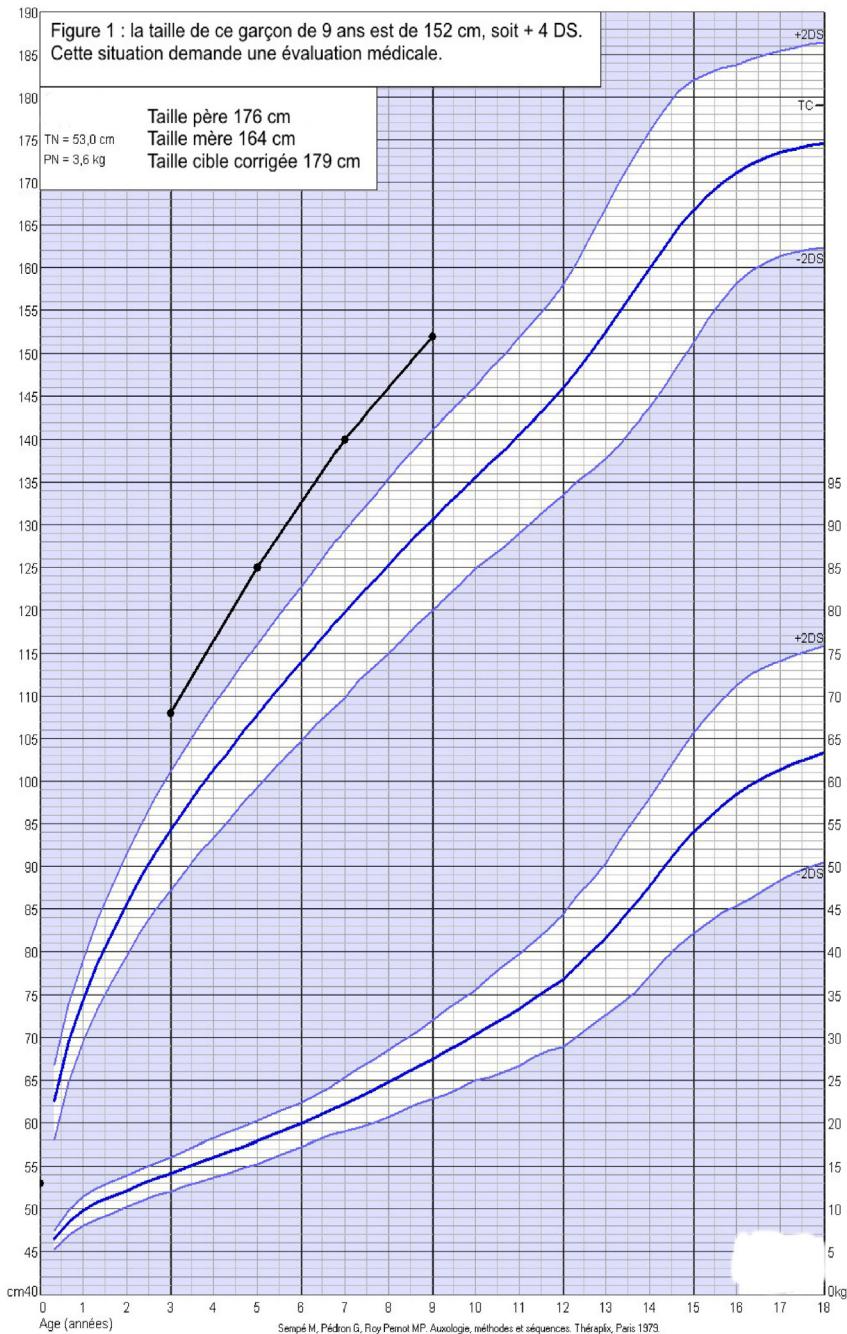


Fig. 1. Size >+2 SD of the average population for age and sex, and >+2 SD the target genetic size.

Although the definition of tall stature is statistical (and therefore affects almost 2.5% of the population), it is a much less frequent reason for consultation in Paediatrics than being small (10 to 20 times less frequent), because tall stature is often perceived as beneficial.

The request is often the result of a bad psychological tolerance towards tall stature, more often observed among girls [1–3]: manifestations of anxiety and depression are more frequent among girls of tall stature and could persist in adulthood [4]. However, women of tall stature have also reported the perception

of increased professional authority, accompanied by difficulties with male subordinates, but no social or family disadvantages.

1.2. Initial management

The consultation will establish the height-weight growth curve, body mass index, and cranial perimeter of the child, collect the term and birth data (weight, length, and head circumference), the height of the parents and first degree relatives (and second degree), pubertal timing of the parents (age of menarche

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