Current topic**The disability of short stature**

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Few paediatricians would deny that children with extreme short stature face a disability that may affect their physical, psychological, and social well being. Though considerable research has been performed on the nature of this disability, it has been focused on patients who are very short, secondary to conditions such as growth hormone deficiency or achondroplasia, and are attending specialised clinics. Increasingly, however, children are being referred who represent the extremes of normal variation in growth and physical development. They do not have a pathological condition and yet their parents are seeking medical advice, usually at their own instigation or at the suggestion of their family doctor, friends, or relatives. Though some individuals, such as Napoleon, are said to have been spurred on to greatness (or infamy!) by their small size, it is widely assumed that short stature, even within the normal range for the population, represents a disability and with the recent availability of treatment with biosynthetic growth hormone this assumption requires further study.

This review will examine the evidence available on the disability of short stature, which to date is based almost entirely on 'pathological' short stature. It will then summarise the relevance of these studies to 'short normal' children.

Studies in children with extreme short stature

A variety of diagnoses are represented, of which growth hormone deficiency is the commonest. In all the studies, however, short stature was the main medical problem.

Psychological/social features. Being unusually short would seem to have an adverse effect on psychological and social well being. High incidences of behavioural disorders have been found^{1 2 3} and particular characteristics identified. Of these, social isolation and lack of appropriate aggressive drive are perhaps the most prominent.^{2 4-7} Usually, the

level of interaction with children is determined by assessment of their age based on their physical appearance; in short children, age tends to be underestimated, and this can lead to the child being 'infantilised' by parents, peers, and teachers.^{1 4} In response the child may adopt babyish behaviour or become a 'mascot' figure.^{2 8} Depression,^{1 8} low self esteem,^{4 5 7} poor social coping,⁶ and an increased incidence of somatic complaints⁵ are also described.

Though adolescents may fare worse than younger children, the published works are inconsistent about the effect of sex on the problems of short stature. This may partly reflect the small numbers and the variety of diagnoses within and between studies as growth hormone deficiency is commoner in boys and Turner's syndrome occurs only in girls. Some studies have shown a higher incidence of personality or behavioural disturbances in adolescent girls,^{6 9} while a psychoanalytic approach suggested that problems might be worse in adolescent boys,^{8 10} the latter being supported by clinical referral patterns. Abbott *et al* found no differences in emotional functioning between the sexes.¹

Intellectual function. In some conditions in which short stature is a part, such as fetal alcohol syndrome, low intelligence quotient (IQ) and retarded development are described features. In most cases of short stature, however, distribution of IQ is within the normal range, and mean scores for study groups approximate to that of the general population.^{1 3 6 11} Some authors have found lower scores on testing particular areas—for example, visual motor integration¹ and spatial orientation,⁶ but others have noted no differences in subscores.^{3 11} The heterogeneity of the diagnostic groups might have accounted for the high incidence of low IQs in a few small samples and Turner's syndrome is associated with poorer non-verbal performance and difficulty in spatial orientation, which is likely to affect samples containing this diagnosis.

School achievement. Early studies showed scholastic underachievement with discrepancy between IQ and academic functioning.³ Steinhausen and Stahnke⁶ noted, however, that IQ and grade achievement were similar in their case and control groups but that children with short stature of unknown aetiology were more likely to be at a lower grade than expected for their age. In another small sample Abbott *et al* found that five of 11 children with growth hormone deficiency had been retained at least one grade but that in four the reason given was absenteeism caused by admission to hospital and poor academic achievement was usually associated with low IQ.¹

A short longitudinal study of school achievement has been performed recently. In 1982 the achievement of 56 children attending a growth clinic for short stature of varying aetiology was assessed. All children had at least average IQ, but 32% had repeated a grade and their parents usually cited immaturity and small size as the reasons, with no reports of scholastic underachievement. Two years later (1984) the children who had repeated a grade (adolescent girls and young boys were most likely to have been retained) were retested; they still showed lower academic achievement and were functioning at a less than appropriate level for their current grade, despite their retention. Specific questioning of parents now revealed that they could accurately assess their child's academic abilities yet had previously (1982 survey) blamed the problems of short stature for grade retention.^{9 12}

School functioning is also affected by interaction with teachers and peers. Short children may be subject to special treatment at school, often being overprotected and treated as if younger than their actual age, and this in turn can lead to underachievement.¹³ In the studies of Holmes *et al* described above teachers and parents thought that short children had appreciable adjustment problems, and in the group that had repeated a grade teachers (but not parents) thought there were more personality and behavioural disorders.^{9 12}

Once again, the heterogeneous nature of the small groups studied may confuse the issue as many patients had either growth hormone deficiency (which may be associated with central nervous system disorders) or short stature of unknown aetiology (many syndromes involving retarded development are also associated with short stature). Normal intelligence but scholastic underachievement, however, is a recurring theme.

Adult attitudes. The way in which parents perceive and subsequently handle the problems of their short children is critical to the child's own adjustment.

Denial of the child's problems has been noted, with overestimation of his present and future height;^{1 4 8} this may lead to severe disappointment with response to treatment with growth hormone.¹⁴ Parents often thought that their child wished to be 'babied', but children's adjustment and maturity were better if they had been treated by their parents at an age appropriate level.^{2 4} Commonly perceived problems were teasing by peers^{1 9} and a younger sibling overtaking the child in height.^{1 8} Perhaps as a consequence, parents sometimes exhibited over-protective or compensatorily lenient behaviour, while possibly reinforcing the infantilism by their own actions.^{4 5}

Interaction with other adults is also important and the perceptions of teachers have been mentioned previously. The tendency to treat children at a size appropriate rather than age appropriate level is emphasised by the study of Rotnem *et al*, in which it was noted that specialist medical and nursing staff involuntarily treated the children with growth hormone deficiency as if they were younger than their actual age, despite theoretical awareness of their problems.⁴

Studies in adults

Like studies in children, observations on adults have been made almost exclusively on clinic attenders with extreme short stature, usually secondary to bony dysplasias (especially achondroplasia) and growth hormone deficiency.

Various problem areas have been identified among adults with achondroplasia. Marriage was less common than in the general population and if achieved was sometimes associated with other problems such as minor psychiatric disorders, suggesting that it was not always a supportive relationship. Although employment was generally found, many thought that they had had to overcome considerable prejudice to obtain and maintain their jobs. Those with achondroplasia felt stigmatised but employed various socially compliant coping mechanisms, such as humour, to deal with this, and few personality differences were found between them and standard populations. Where they existed they were usually 'acceptable' traits, such as increased extraversion and conformity, and scores for anxiety, neuroses, and hostility were normal.^{15 16}

Adults with growth hormone deficiency showed similar characteristics to those with achondroplasia in the sample of Brust *et al*,¹⁵ but Clopper *et al* found an increased incidence of abnormalities.¹⁷ Adults with growth hormone deficiency had inertia of libido, and deficient socialising and pair bonding behaviour, with depression and other behavioural

disturbances being much more prominent than in those with achondroplasia. Canadian adults with growth hormone deficiency who had received growth hormone as children had normal educational attainment but lower rates of marriage and employment than the general population and were socially isolated. These disadvantages were not clearly related to the response to treatment.¹⁸

Psychological effects in children at the extremes of normal variation

So far the evidence presented had considered patients who are so short that their stature could be considered pathological. A much larger group consists of those individuals whose growth patterns represent the extremes of a wide normal variation. Little research has been conducted in this population, with the notable exception of the studies of Mussen, Jones, and Bayley.¹⁹⁻²¹ Though their data was collected in the 1930s and '50s, they are interesting because of the long follow up period into adult life. They took a sample of adolescents attending a normal school and selected the 20% who were earliest developing in puberty and the 20% who were latest, following them longitudinally from 12 to 17 years. The two groups were also recontacted when they were in their early 30s.¹⁹⁻²¹

The selected difference between these two groups was the tempo of puberty, but as this has a direct effect on height the late maturing group had heights in the lower part of the normal range (10th-25th centiles) and the early maturing group in the upper part (60th-90th centiles). The results showed considerable differences between the two groups. Late maturers had high scores on measures of negative self conceptions, inadequacy, dependence, rejection, and disturbed relationships with parents. Early maturers scored high on self confidence, independence, and dominance. Trained observers also rated the groups differently; they judged early developers to be more physically attractive, relaxed, matter of fact, and less affected and late maturers to be more expressive and eager (often in a childish way), tense, and with attention seeking mannerisms (though this may have been as much a difference in the observers' attitudes as in the boys' behaviour).

Many subjects were contacted again when they were in their early 30s, by which time the differences in their physical appearance were extremely small (height difference ½ inch). The two groups were found to be very similar in their social characteristics—for example, marriage, children, income, education—but the psychological differences noted during adolescence had essentially persisted into adult life. Those who had been early maturers

scored high on socialisation dominance, self control, and responsibility, while the late maturers were less dominant, wished for more support, and tended to be rebellious but non-assertive.

Implications for growth hormone treatment

The handicap associated with extreme short stature led to vigorous attempts to find a suitable treatment. The isolation and preparation of human growth hormone provided hope for children with growth hormone deficiency but was not thought to be useful in other conditions or in normal short children. Extraction from pituitary glands severely limited supplies and few clinical trials could be carried out. In the past few years, however, small series have suggested that growth hormone might be used to increase growth in normal short children and that up to 50% of children with genetic short stature or constitutional growth delay might respond to treatment.²²

There remain several unknown factors. Firstly, availability of human growth hormone has limited trials to low dose regimens and there is virtually no data on the effects or side effects of higher doses. Secondly, practical considerations have limited duration of treatment (usually 6-12 months) and so we do not know how long treatment should continue. Thirdly, no prospective method of distinguishing responders from non-responders has been found. Lastly, the crucial issue, the effect of treatment on final adult height cannot be addressed for many years. The question of who should have growth hormone has been reviewed recently²² and discussed by an ad hoc committee of paediatric endocrinologists representing the American Academy of Pediatrics. The committee maintained that the only established indication for treatment with growth hormone was growth hormone deficiency and that its use in all other cases must be considered experimental. Furthermore, they considered that the handicap associated with short stature might be due to underlying emotional disturbances and so might not be relieved by treatment with growth hormone and concluded 'if it ain't broke, don't fix it'.²³ The prevailing negative attitude to short stature in Western societies and the ethics and philosophy of treatment with growth hormone in short normal children was also extensively discussed in a Hastings Center report three years ago.²⁴

The recent development of commercially viable biosynthetic growth hormone products means that the supply of growth hormone is theoretically unlimited, although the cost of the preparation is

high. This paves the way for increased clinical use but also potential abuse, both of which were anticipated and discussed at an international conference held in 1983.²⁵ It was expected that paediatricians would come under parental pressure to prescribe growth hormone and one consensus opinion was that 'much of that pressure will come from parents whose children are not fulfilling parental expectations in competitive sports, social interactions, and academic achievement.'

While to be very short is clearly disabling psychosocially as well as physically, we know much less of the effect of having a growth pattern at the extreme of the normal range. An increasing number of patients seeking specialist medical advice for short stature, however, belong to this latter group; presumably they feel disabled and many are anxious to have treatment that will make them taller. Aside from the diagnostic difficulties²² and therapeutic uncertainties, the nature of this disability poses additional questions. Are these children experiencing an abnormal psychological reaction to a physiological growth pattern? If so, they are probably not representative of the short population at large and might be helped by counselling in addition to or instead of treatment with growth hormone, particularly as it will be impossible to guarantee a favourable outcome from treatment with growth hormone. Psychological disturbances may be created as well as alleviated by treatment. Overexpectation of treatment by both parents and their children with growth hormone deficiency has led to great disappointment and feelings of failure, despite a medically acceptable increase in growth rate.^{7 10 14} Some children have found adjustment to the unusual experience of growing difficult, with aggression and depression becoming prominent, while pre-existing traits of low self esteem and depression were not relieved and were sometimes aggravated by treatment.^{2 7} In the Canadian series 21% of patients needed psychiatric or psychological counselling during treatment with growth hormone.¹⁸

An alternative answer may be that these normal short children seeking medical advice represent a vast number in the community, whose short stature is disabling but who, for various reasons, have not sought or had access to medical care. Included in this number may be children who are short secondary to another medical problem, such as cystic fibrosis or renal disease. We do not know how much of an additional burden this places on them or of what benefit treatment with growth hormone may be. In this alternative scenario the large numbers of children who may be helped by treatment with growth hormone emphasise still further that its careful allocation and evaluation is essential for the

benefit of the individual child and the whole community.

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