SHORT REPORT

Growth study of cri du chat syndrome

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Abstract

We compared the growth of children with cri du chat (5p-) syndrome with the 1990 UK growth curves. Most subjects had impaired growth, particularly of head circumference. The more emaciated the child the more pronounced the microcephaly, showing the need for growth and nutrition monitoring.

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Impaired growth is common in cri du chat (5p-) syndrome (CDCS) and, with international collaboration, syndrome specific growth charts have been published.¹ The skewed distribution of weight in CDCS towards underweight must be considered when using these charts, and because of differences between ethnic populations it is important that they are used in conjunction with reference data for the general population of a child's country of origin.

The loss of genes in CDCS may influence growth directly, through gene products modifying metabolic pathways, and indirectly, because of feeding difficulties resulting in malnutrition. Feeding problems are common in CDCS and parents reported receiving little or no help from professionals with regard to feeding problems extending beyond early infancy.²

We aimed to compare the anthropometric measurements of British Isles white children with CDCS with the cross sectional stature, weight, body mass index, and head circumference reference curves for the UK, 1990.^{3,4}

This cross sectional comparative study is part of a larger nutritional and anthropometric project begun in 1995, which received ethical approval from the Research Ethical Committee of the University of Ulster.

Methods

Subjects were recruited through the Cri du Chat Syndrome Support Group. All measurements were taken by trained professionals. The average of three measurements was recorded. Body mass index (BMI) was calculated.

Data were analysed using the 1996 revision of the Child Growth Foundation's growth reference program, which allows the conversion of

Table 1 Means and standard deviations of BMI, height, weight, and head circumference standard deviation scores for all subjects

Age group	n	Mean	Standard deviation	Range
<i>Body mass index for age SD</i>				
0 to 4.99 years	31	-1.59	2.29	-6.47 to 4.53
5 to 9.99 years	15	-1.58	1.67	-5.37 to -0.96
10 years and over	9	-0.54	1.94	-4.31 to 1.92
Whole group	55	-1.40	2.80	-6.47 to 4.53
<i>Height for age SD</i>				
0 to 4.99 years	31	-1.24	1.6	-3.79 to 2.16
5 to 9.99 years	16	-2.73	2.02	-7.43 to -0.22
10 years and over	9	-1.69	1.21	-4.13 to -0.54
Whole group	56	-1.74	1.77	-7.43 to 2.16
<i>Weight for age SD</i>				
0 to 4.99 years	32	-2.04	1.82	-5.54 to 1.28
5 to 9.99 years	15	-2.81	2.25	-8.45 to -0.05
10 years and over	9	-1.39	2.05	-5.42 to 0.96
Whole group	56	-2.14	2.00	-8.45 to 1.28
<i>Mid head circumference for age SD</i>				
0 to 4.99 years	32	-4.27	1.67	-8.75 to -0.85
5 to 9.99 years	16	-4.16	3.19	-9.48 to 2.25
10 years and over	9	-3.02	2.03	-7.24 to 0.27
Whole group	57	-4.00	2.25	-9.48 to 2.25

each measurement to a standard deviation score (z score).

Results

There were 30 girls and 27 boys (age range 0.5-16.67 years, mean 5.55, SD 4.18) with CDCS in the sample, approximately 38% of the cohort aged under 23 years known to the support group in September 1999. Table 1 shows results of growth measurements. Gender differences in standard deviation scores were not statistically significant. There was a statistically significant correlation between mid head circumference for age and BMI for age ($r = 0.56$, $p < 0.01$, one tailed test).

Discussion and conclusions

The wide range of standard deviation scores, and particularly the negative standard deviation scores, for each of the growth parameters shows that it is clinically relevant to assess British Isles children with CDCS using the UK 1990 centile charts. Most subjects were microcephalic, small and light for age, and the more emaciated the child the more pronounced the microcephaly.

BMI scores suggest that most children were lean, rather than emaciated, but this may reflect

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relatively good muscle development.²⁻⁵ Insufficient cross sectional data were available to ascertain either age of peak height velocity or BMI rebound.

We believe these results raise the question of whether some growth impaired children with CDCS suffer from undernutrition, and highlight the need for growth and nutrition monitoring.

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obtained through the Child Growth Foundation, 2 Mayfield Avenue, London W4 1PW, UK.

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