A new chart for weight control in Duchenne muscular dystrophy

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SUMMARY Weight control is desirable in the muscle wasting conditions. A new chart is presented to allow the prediction of an ideal weight, free of excess fat, specifically for boys with Duchenne muscular dystrophy.

Patients suffering from muscular dystrophy often put on weight as the disease progresses. It is not uncommon for boys with Duchenne muscular dystrophy to have weight values greater than the 90th centile on normal growth and development charts, despite having a smaller muscle mass than normal boys of the same age. It has been shown that therapeutic weight reduction is possible in these patients, and loss of unnecessary fat lessens the burden of weakened muscles and aids mobility.

Prevention of excessive weight gain in the younger patient is preferable to severe dietary restriction in the already obese subject. Normal growth and development charts make no allowance for the progressive loss of muscle that is occurring as the muscular dystrophy progresses throughout childhood. A child with pronounced muscle wasting looks 'cachectic' without the excess fat filling out his body contours, something parents find hard to accept. The loss of muscle coupled with reduced activity implies accumulation of fat tissue if weight gain approximates that of normal growth charts.

Method

The 24 hour excretion of creatinine in the urine as a breakdown product of phosphocreatine in muscle, assuming 1 mmol creatinine equals 2.625 kg of muscle, provides a reliable estimate of the contribution of muscle to body weight in the absence of renal disease.  

Theoretical weight centiles for zero muscle mass can be drawn from data on normal creatinine excretion and from standard weight and height centile charts. From a knowledge of the patient's creatinine excretion the muscle bulk may be derived and added to the zero muscle weight chart to produce an ideal weight.

Results

A new chart is presented, which gives ideal weight guidelines for weight control in boys with Duchenne muscular dystrophy (figure). From observations of body composition and creatinine excretion in boys with Duchenne muscular dystrophy we have found a close relationship between the reduction in total body potassium and that of 24 hour urinary creatinine excretion. Both total body potassium and urinary creatinine excretion, when related to the predicted values for the individual's height, declined with age at an average rate of 4% per year from 6 to 17 years. A similar 4% per year loss in muscle strength has been observed in boys with Duchenne muscular dystrophy. At 6 years a boy with Duchenne muscular dystrophy has only 60% of his predicted muscle mass, and at 16 years only 20%. The reduction in creatinine excretion can be calculated and from this the muscle bulk can be derived and added to the zero muscle weight charts. Based on this assumed 4% per year decline in muscle mass the 90th, 50th, and 10th 'ideal' weight centiles have been calculated to allow for progressive loss of muscle.

Discussion

This chart can be used in the normal way by selecting the appropriate height related centile. This chart is only applicable for Duchenne muscular dystrophy. In other muscle wasting conditions
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Figure. Ideal weight centile chart for boys with Duchenne muscular dystrophy. Based on data from Edwards et al., Edmonds et al., and Tanner and Whitehouse. Assumes a 4% per year decline in muscle bulk.
measurement of 24 hour creatinine is advised with calculation of the ideal weight from the zero muscle chart. To obtain a practical objective for dieting allowance must be made for the increased fibrous tissue accumulation that takes place in the dystrophic disease process.

How to use the chart

This chart is used in conjunction with the Tanner and Whitehouse longitudinal standards for height and weight.

1. Find appropriate height centile.
2. Predicted ‘ideal’ weight is read off the chart using the height centile line.
3. A practical objective for diet control is achieved by adding to the ideal weight an extra kg for every year the boy is over 10 years—for example, at 15 years this would be 5 kg extra.

The support of the Muscular Dystrophy Group of Great Britain and Northern Ireland is gratefully acknowledged.

Does ABO incompatibility matter?

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SUMMARY The incidence and severity (peak serum bilirubin concentration) of clinically detectable jaundice was determined retrospectively in 110 elution positive cases of ABO incompatibility. Neither the incidence nor the severity of jaundice in the study group differed significantly from a control group. In the individual case Coombs positivity and/or a strongly positive elution test may be a helpful predictor of jaundice occurring but not of its severity.

With the declining incidence of Rhesus disease, ABO incompatibility is said to be the commonest cause of haemolytic jaundice in the newborn. This is not surprising in view of the fact that 20–25% of pregnancies are ABO incompatible. Compared with Rhesus incompatibility, however, the jaundice is usually mild and characteristically later in onset. Furthermore, associated anaemia rarely occurs. These observations prompted us to consider the clinical importance of ABO incompatibility by a retrospective case-control study.

Patients and methods

In this hospital cord blood is routinely taken from the infants of all group O mothers. This is tested for blood group and presence of antibodies by an elution test and direct Coombs test. The elution test is performed by washing the infant’s red cells twice in saline followed by heating to 56°C to dissociate the maternal antibody from the antigenic sites. The resultant solution is centrifuged and the supernatant (eluate) tested against adult red blood cells of known group. Agglutination of the cells confirms the presence of specific antibody. The elution test detects maternal anti-A or anti-B haemolsins (IgG antibodies) on the infant’s red blood cells.

Routine screening was instituted in 1978 after reports that it reduced the need for exchange transfusions in ABO incompatible infants by allowing early detection. Data were collected on 110 ABO incompatible deliveries (94 mother group O/infant group A (O-A), 16 mother group O/infant group B (O-B)) in which the cord blood elution test was positive. The elution test was graded as weakly positive or strongly positive. A control group