ESTABLISHMENT OF BONE DENSITY STANDARD CURVES FOR CHILDREN WITH TYPES III AND IV OI SHOWS DIFFERENT PROGRESSION PATTERNS FOR THE TWO TYPES.

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Purpose: Establishment of bone density standard curves for children with osteogenesis imperfecta is essential for determinations of the relative severity of osteoporosis of OI children. Furthermore, the dependence of bone density measurements on the type of OI, the age and height of the child are unknown.

Methods: Lumbar (L1-L4) bone density measurements were obtained by DEXA using a Hologic QDR 2000 densitometer. Both single beam and array beam (high resolution) acquisition was performed. Raw data was analyzed using low density software supplied by the manufacturer. Univariate and multivariate regression analyses were done for all measurements, grouped by OI type. Multiple measurements of the same child were handled as having less dependence with increasing interval between values.

Results: We analyzed bone density measurements made on 35 OI children who were regular participants in the NICHD OI Program, including 16 type III and 19 type IV OI. Type III children ranged from 1.5 to 16 yrs old at time of BD measurement. They contributed 85 single bean and 48 array beam measurements. Type IV children ranged from 2.5 to 19 yrs old at time of BD measurement. They contributed 130 single beam and 54 array beam measurements.
For bone density measurements by single beam acquisition, the increase in bone density is predominantly determined by age with some contribution of height in type IV OI. For univariate analysis:

Type III BD = .139 + Age(.0075)
Type IV BD = .165 + Age(.0161)
When single beam measurements are converted to z scores, we obtain a parabolic curve with respect to age for both OI types. Z scores become less negative until age 10-11 yrs in Type IV and age 8-9 yrs in Type III, after which they fall again.

Conclusions: The clinical phenotype of types III and IV OI have different patterns of development of single beam lumbar BD. The slopes of the mean for single beam values were significantly different ($p = .03$) when analyzed by a normal z test. Comparison of predicted values for all children shows that the distinction of types III and IV is highly significant ($p < .001$) over age 5yrs. Analysis of BD measurements converted to z scores demonstrates that there is a greater response of type IV BD to puberty but this response is proportionately less than the pubertal BD increase of unaffected children. Establishment of these standardized curves will facilitate comparisons of lumbar bone density and the spinal symptoms of OI, vertebral collapse and scoliosis.