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[1 Letter to the Editor]

Optimizing bone health in cerebral palsy across the lifespan

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SIR–We read with interest the new osteoporosis care pathway from the American Academy of Cerebral Palsy and Developmental Medicine (AACPDM), highlighting the important issue of bone health in children with cerebral palsy (CP).[1] The focus of the guidelines is on optimizing nutrition, vitamin D supplementation, encouraging weight-bearing to promote skeletal development, and considering bisphosphonate administration as a preventive and therapeutic measure. We welcome the timely release of the guidelines, given that individuals with CP are living

longer and are likely to develop osteoporosis as a consequence of factors beginning in childhood.

With recent studies documenting increased skeletal fragility in adulthood, we would like to raise a number of points that may impact the assessment and management of bone health in childhood and transition to adulthood.

GONADAL STATUS

Puberty plays a major role in accrual of bone strength in growing children and adolescents, so that delayed or disordered pubertal development may have detrimental effects.

In young adults with CP, we found that 20% were hypogonadal on clinical and biochemical assessment.[2] Adults with hypogonadism had lower lumbar spine bone density and lower lean tissue mass on dual-energy x-ray absorptiometry (DXA) assessment than eugonadal adults with CP. The majority had hypogonadotrophic hypogonadism, with possible explanations including poor nutrition and chronic illness likely leading to functional hypothalamic hypogonadism. Alternatively, hypothalamic-pituitary axis dysfunction may result from the early injury to the immature brain seen in CP. It is unclear when hypogonadism is manifesting in CP; it may be during late childhood, given a cross-sectional study of 207 children with CP by Worley et al.[3] which demonstrated altered pubertal progression. Timing of puberty will also be influenced by nutritional status, with overweight promoting earlier onset and underweight delaying progression.

Given the negative effect of established hypogonadism on bone mineral density, clinical assessment of pubertal progression with Tanner staging is recommended. The onset of breast development marks the onset of puberty in young females; increase in testicular volume to 4ml marks puberty onset in young males. Puberty is delayed if pubertal change has not begun by age 13 in young females; by age 14 in young males. If there is concern regarding pubertal delay, further investigations

should include bone age x-ray with biochemistry for gonadotrophins and testosterone/estradiol. Consideration of sex hormone replacement should be made if deficits are identified, bearing in mind that pubertal growth, psychosocial sexual development, and menarche are recognized as challenging for caregivers, and concerns about aberrant pubertal progression may not be raised with health care professionals.

BONE DENSITY MEASUREMENTS

In the recent guidelines by Ozel et al., the lateral distal femur or the total body is the site of choice for bone density measurement using DXA.[1] The distal femur is clinically relevant, being a common site of fracture in children, and has established reference ranges in children. It has been validated with a correlation noted between z-scores and fracture history in children with CP.

In adults however, there is no normative data for lateral distal femoral assessment and this technique is not readily available. A number of studies in adults with CP have used the lumbar spine, femoral neck, or total hip for measurement,[2,4,5] with z-scores at these sites correlated to fracture risk. Furthermore, this site may be more relevant to the more typical fragility fractures seen in adults with CP, which include vertebral, ankle and rib fractures. In adolescents transitioning to adult care, it may be worth considering using these sites in addition to the distal lateral femur to prepare for ongoing monitoring in adulthood.

BODY COMPOSITION

The use of whole body DXA scanning also allows the assessment of body composition. This is an invaluable tool which allows differentiation between fat and lean tissue mass, with relevance to those caring for patients with nutritional concerns. Nutritional deficits can lead to poor muscle mass and failure of skeletal growth. Given that body composition can be calculated from a whole body scan with no further radiation

exposure, it is worthwhile requesting this assessment in addition to bone density.

TRANSITION TO ADULT CARE

In summary, osteoporosis in CP is a disease that begins in childhood but manifests throughout life as fractures. Ensuring follow-up is paramount in maintaining quality of life and mobility of those who have had previous fragility fractures, as well as those with multiple risk factors for reduced bone density. This is a shared task for both paediatricians and adult physicians.

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