

Severe Infantile Cholestasis as a Cause of Silent Rib Fractures

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Clinical Image

We cared for a 7 month old girl with history of extrahepatic biliary atresia (EHBA) and immediately underwent hepatoportoenterostomy (HPE) at around age 2 months of age. On physical examination, she was noted to have icteric sclerae, generalized jaundice, hepatosplenomegaly, and rachitic rosary on the chest wall (Figure 1).

Her vitamin D levels consistently were below 15 ng/mL for several weeks after HPE despite increments of oral dose of cholecalciferol or ergocalciferol from 1200 to 8000 IU/day. Due to vitamin E deficiency, Aquasol E® (TPGS or water soluble form) was co-administered with vitamin D dosing. Figure 2 demonstrates bilirubin profiles and 25-hydroxy vitamin D concentrations over time after HPE.

Despite biliary drainage, she had significant cholestasis, ascites, malnutrition, and transient melena. Subsequently she was evaluated for a liver transplantation. A chest X-ray and an abdominal CT scan were obtained with an incidental finding of healing right sixth and seventh lateral rib fractures with callus formation and healing left seventh lateral rib fracture with callus formation (Figures 3 and 4).

There was no previous history of known trauma, and there were no social concerns regarding this child. She had no signs or symptoms to suggest pain or irritability following HPE. A skeletal survey was obtained that did not reveal any additional fractures, however, there was evidence of mild underlying demineralization which confirmed our suspicion that this incidental bone fracture finding was secondary to rickets.



Figure 1: Rachitic rosary on the chest wall.

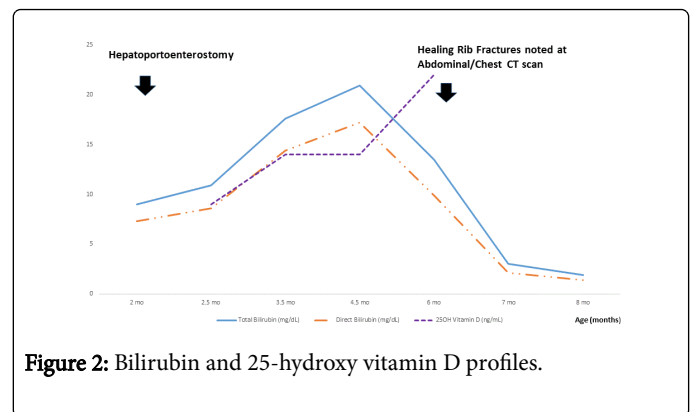


Figure 2: Bilirubin and 25-hydroxy vitamin D profiles.

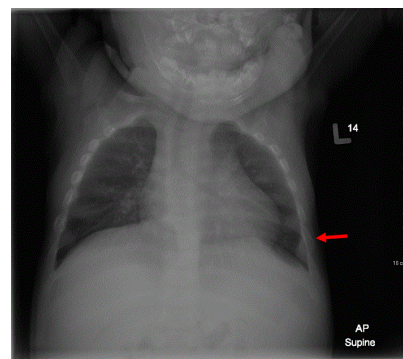


Figure 3: CT scan showing sixth and seventh lateral rib fractures along with callus formation left lateral 7th rib (CXR).

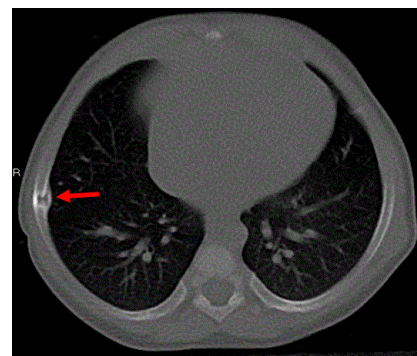


Figure 4: Callus formation at left lateral 7th rib.

EHBA is a progressive cholangiopathy that results in fibrotic obliteration of intra and extrahepatic bile ducts in infants significant from the first 2 months of life [1,2]. Osteopenia and rickets are a known complication of EHBA, regardless of a successful outcome after HPE [3]. Absorption of ingested vitamin D is significantly impaired in children with cholestatic liver disease such as EHBA [1]. Several fat soluble vitamin deficiency, particularly vitamin D deficiency was prevalent and inversely correlated to serum total bilirubin levels (TB>2 mg/d) [1]. Bone fracture secondary to rickets can be highly suspicious for non-accidental trauma and clinically missed even when a high dose of vitamin D is administered. Glasgow et al reported the radiographic onset of osteopenia at about 12 months of age in infants with EHBA. [4] Bone fractures in children <1 year of age with child abuse involves rather long bones especially the metaphyses and posterior ribs [5,6]. Global imaging of the bones in this child revealed generalized osteopenia on a skeletal survey but no pathologic fracture of any long bones or posterior ribs which are common suspicion of non-accidental trauma such as child abuse.

Infants with persistent cholestasis (serum TB>2 mg/dl) remain at a higher risk for rickets despite early appropriate supplementation of vitamin D [1,2]. Prompt treatment for vitamin D deficiency is warranted in infants with EHBA. Close monitoring of vitamin D and other fat soluble vitamins is essential in the care for children with cholestatic liver disease, particularly EHBA. A more water soluble form of vitamins (TPGS) is preferred or co-administration of fat soluble

vitamin such as vitamin D with available TPGS form of other fat soluble vitamin such as Aquasol E® is practical [1]. In summary, osteopenia described as cortical thinning and trabecular bone loss on an X-ray, findings consistent with rickets, and/or multiple fractures would EHBA and severe vitamin D deficiency rather than child abuse as an etiology of bone fractures in this case.

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