

## Poster

### **Schneckenbecken dysplasia in fetus: A rare form of familial osteochondrodysplasia Report of 4 cases**

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Schneckenbecken dysplasia is a rare lethal osteochondrodysplasia with autosomal recessive mode of inheritance. It's a part a large spectrum of lethal short -limbed skeletal dysplasia with platyspondylia .

We describe 4 cases of Schneckenbecken dysplasia occurring in 3 tunisian families ( 2 cases in one family). Theses fetus were issued from consanguineous marriage. Ultrasound diagnosis of fetal short limbs was done at 20, 22, 17, and 21 weeks of gestation.

Fetopathological examinations associate to skeletal radiographics were performed . they showed a typical feature characterized by small ilia with medial snail-like projection. Chondro-osseous morphology showed abnormal vascularization and increase of chondrocytes with large nuclei in the epiphyseal region of long bone with normal ossification palate.

We discuss than the diagnosis criteria and the genetic counseling of this type of chondrodysplasia and we review the protocol of bone investigation in case of fetus involved by osteochondrodysplasia.

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