

Pseudoarthrosis of the tibia congenita

Congenital tibiae congenita (pseudoarthrosis tibiae congenita) (*Congenital tibiae congenita*) is a serious congenital defect and belongs to the most difficult problems in pediatric orthopedics. It is a unilateral disorder of the normal bone structure of the distal half of the tibia , sometimes also of the fibula . It is not related to skeletal fractures that occur as a complication of fractures .

Etiology

The cause is defective differentiation of the mesodermal structures of the limb during embryonic development. It is often found in patients with neurofibromatosis .neurofibromatosis.

Clinical picture

Children are usually born with a lower leg joint, or it can occur as a result of a pathological fracture in the 1st year of age. Typically, there is a distinct angulation of the lower leg, going as far as varositis and antecurvation, in contrast to congenital kyphoscoliosis of the lower leg, when we find retrocurvation and valgosis. Pathological mobility is observed.

X-ray image

There are various classifications, the most used is according to **Boyd**:

- **I.** – predominance of anterior angulation of the tibia (already at birth), simultaneously with other congenital defects;
- **II.** – at birth, the patellar joint is absent, the tibia is angulated and narrowed in the shape of an hourglass, the fracture occurs during the first years of life, the patellar joint is usually associated with neurofibromatosis;
- **III.** – cystically changed tibia, anterior angulation;
- **IV.** – the hip joint arises as a fatigue fracture in the sclerotized tibia;
- **V.** – dysplasia of the fibula, the joint develops on the fibula, it can also be present on the tibia;
- **VI.** – the finding corresponds to a neurofibroma or schwannoma , the joint may not develop.

Therapy

The treatment of an already formed hip joint is always surgical and very difficult, as bone healing in pathological terrain takes place differently. The options for surgery are: bridging the defect with a bone graft, so-called spongioplasty, ntramedullary fixation, so-called osteosynthesis,, external fixation, osteosynthesis with a plate and bridging with a vascularized autogenous fibular transplant - osteoperiosteal graft.

Odkazy

Použitá literatura

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Kategorie:Ortopedie Kategorie:Pediatric Kategorie:Genetics Kategorie:Významně pozměněné zkontrolované články