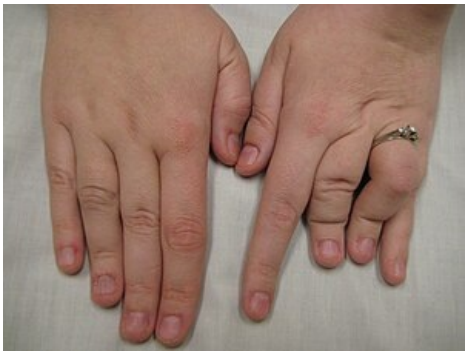
Larsen syndrome

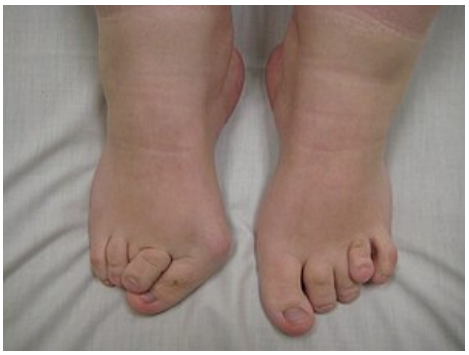
Larsen syndrome is a very rare syndrome with hyperlaxity (hypermobility) of joints and ligaments characterized immediately after birth:

- ***face changes*** – flattening of the face, widened and flattened root of the nose, wide-set eyes, hypertelorism
- ***multiple joint dislocations*** – mainly dislocation of the knee joints
- ***cervical hyperkyphosis*** – there is a risk of damage to the spinal cord

Larsen syndrome



Hands of a patient with Larsen syndrome



Legs of a patient with Larsen syndrome

Patogenesis

- generalized mesenchymal disorder
- autosomally inherited form is caused by a mutation of the FLNB gene on chromosome 3 (3p14.3)

Clinical picture

- changes in the face, flattening of the nose
- hyperextension of the lower limbs, rigid bilateral pes equinovarus, hip joints bilaterally luxated and highly mobile (due to significant joint laxity), flexion contracture of the elbow
- typical hand changes – long fingers mount on short metacarpals, distal joint of thumb blade-shaped
- spinal changes – cervical kyphosis with progressive instability, more distally kyphoscoliosis
- congenital defects of the heart, aorta, trachea, larynx
- normal intelligence

Therapy

- prevent cervical spinal cord compression by posterior fusion
- surgical (or conservative) treatment of luxation of knee joints, hip joints and finally pes equinovarus

Links

Related articles

- Achondroplasia
- Diastrophic dwarfism
- Thanatophoric dwarfism
- Congenital limb defects
- Developmental hip dysplasia
- Pes equinovarus congenitus
- Congenital developmental defects

Used literature

- DUNGL, P.. *Ortopedie.* 1. edition. Grada Publishing, 2005. ISBN 80-247-0550-8.