# Osteodystrophia fibrosa cystica generalisata

**Osteodystrophia fibrosa cystica generalisata** ( *morbus Recklinghausen, primary hyperparathyroidism*) is a disease belonging to the group of acquired systemic diseases of the bone system. **It must be distinguished from morbus von Recklinghausen, which is synonymous with neurofibromatosis - type 1.**

## Etiopathogenesis

It mainly affects women (mainly in the 2nd decade of life). **The cause is an adenoma of the parathyroid glands** , which leads to hyperproduction of PTH , PTH releases phosphates and calcium salts from the skeleton, thereby increasing phosphaturia and increasing calciuria (it increases calcium resorption in the ascending limb of the loop of Henle, but due to high calcium values, hypercalciuria still occurs) . This results in hypophosphatemia and hypercalcemia .

At the same time, there is an increased formation of osteoid (fibrous remodeling of cancellous bone). Cystic destruction of the skeleton and generalized osteoporosis occur . Fractures/infractions with intraosseous hemorrhages occur at the site of significant weakening of the load-bearing parts of the skeleton .

## Clinical picture

**Fatigue** with reduced physical performance, occasional **pain** in the spine and limbs. In the later stage of the disease, minor **limb deformities** , or spontaneous fractures. **Kidney damage** : nephrolithiasis to nephrocalcinosis .

## Laboratory finding

- hypercalcemia
- hypercalciuria
- hypophosphatemia
- hyperphosphaturia

## X-ray image

X-ray examination performed only after the occurrence of a spontaneous fracture (cystic deposits, thinning of the compacts, enlargement of the medullary cavity). A decrease in the thickness of the vertebral bodies, their expansion, and the occurrence of multiple compression fractures are common . Subperiosteal bone reduction, most commonly seen on the middle phalanges of the fingers. Structural changes in the calf are common.

In the advanced stages of the disease, there are multiple angulations and severe deformities of the load-bearing parts of the skeleton. On CT sometimes parathyroid adenoma.

## Therapy

Causal treatment only **surgical** (removal of parathyroid adenoma). Hypercalcemic crises are treated with hydration and adjustment of the mineral economy.

Orthopedic therapy consists in corrective osteotomy of the resulting deformities, possibly in combination with prolongation performances .

## Differential diagnosis

Fibrous dysplasia (Jaffe-Lichtenstein), cortical fibrous defect, juvenile solitary pseudocyst , myeloma . In all of these diseases (with the exception of plasmacytoma) unilocular/monomelic occurrence, but in fibrous dysplasia the bones of almost the entire skeleton are affected .

# Links

## Related Articles

- Metabolic osteopathy
- Osteoporosis
- Osteomalacia
- Rickets

## References


1. ↑Jump up to:a b c d e f g SOSNA, A., P. VAVŘÍK and M. KRBEC, et al. *Basics of orthopedics.* 1st edition. Prague: Triton, 2001. ISBN 80-7254-202-8 .