

# Paget's disease surgery

There are several neurological complications of [Paget's disease](#). They may concern the [medulla](#), the [cauda equina](#), and also the cerebral trunk, the [cranial nerves](#), and the brain. Complications are rare when the great frequency of Paget's disease is considered. But they should be recognized because they constitute an aspect of the disease the treatment of which is usually well tolerated or even negligible. The signs of involvement of the medulla, of the cauda equina, and those which complicate the [basilar impressions](#) usually develop in a progressive and not very spectacular fashion. Nevertheless, they should be recognized as soon as possible so that therapeutic measures may be taken. The results of modern medical treatments, [calcitonin](#), cellulolytic medications, and possibly others are already sufficiently established to justify their use initially. Indications for surgical treatment appear when a check in medical therapy is observed after three months of treatment <sup>1)</sup>.

A case of cauda equina compression by ossification in extradural fat at a level above the site of Paget's disease in a vertebra was described in 1975. Good recovery followed laminectomy and exposure of the dura mater <sup>2)</sup>.

In general, [conservative treatment](#) of [fractures](#) in [Paget's disease](#) are associated with a high rate of delayed union.

The lumbar region is the most commonly affected site within the [spine](#) followed by the [thoracic](#) and [cervical spine](#). Even though the spine is affected very commonly in Paget's disease, malignant degeneration is exceptionally rare. Multilevel monostotic spine involvement due to Paget's disease is very uncommon. An unusual clinico-radiological manifestation of multilevel thoracic Paget's disease with sarcomatous degeneration presenting as a neurosurgical emergency is reported with a pertinent review of the literature by Tan et al. <sup>3)</sup>.

Surgical [indications](#) for spinal Paget's disease:

1. rapid progression: indicating possible malignant change or [spinal instability](#).
2. spinal instability: severe [kyphosis](#) or compromise of [spinal canal](#) by bone fragments from pathologic fracture. Although the collapse is usually gradual, sudden compression may occur.
3. uncertain diagnosis: especially to R/O metastatic disease (osteoblastic lesions)
4. failure to improve with [medications](#)

Surgical considerations

1. profuse [bleeding](#) is common: if significant bleeding would present an unusual problem, treat for as long as feasible pre-op with a [bisphosphonate](#) or [calcitonin](#).

a) use [bone wax](#) to help control bleeding.

b) [hemostasis](#) may be difficult

2. to treat resultant [spinal stenosis](#): decompressive [laminectomy](#) is the standard procedure in the [thoracic region](#) <sup>4)</sup>.

However, if most of the pathology is anterior, consideration should be given to anterior approach.

3. bone is often thickened, and may be fused with obliteration of **interspace** landmarks. A high-speed drill is usually helpful.

4. post-op medical treatment may be necessary to prevent recurrences <sup>5)</sup>.

5. osteogenic sarcoma

a) surgery and chemotherapy are used, cure is less likely than in primary **osteosarcoma** of non-pagetic origin

b) biopsy proven of the scalp requires en-bloc excision of scalp and tumor.

## Surgical outcome

In 65 patients treated with decompressive laminectomy, 55 (85%) had definite but variable degrees of improvement. Patients who had only minimal improvement were often ones with malignant changes. One patient was worse after surgery, and the operative mortality was 7 patients (10%). Survival with malignant degeneration is <5.5 mos after admission <sup>6)</sup>.

## References

<sup>1)</sup>

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<sup>2)</sup>

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<sup>3)</sup>

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<sup>4)</sup> , <sup>6)</sup>

Sadar ES, Walton RJ, Gossman HH. Neurological Dysfunction in Paget's Disease of the Vertebral Column. JNeurosurg. 1972; 37:661-665

<sup>5)</sup>

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