



The spectrum of haemophilic pseudotumours: Management at a single centre in India.



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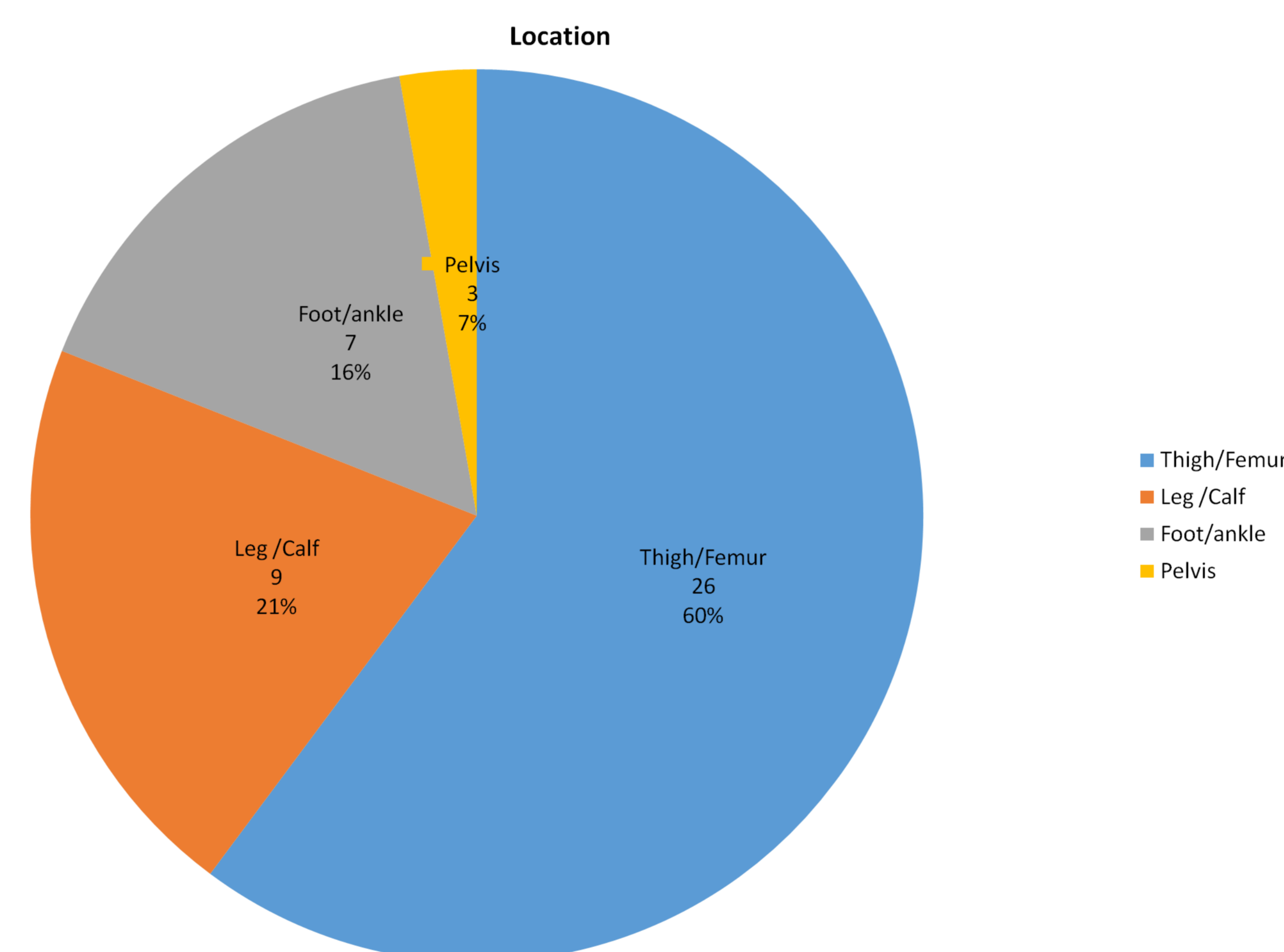
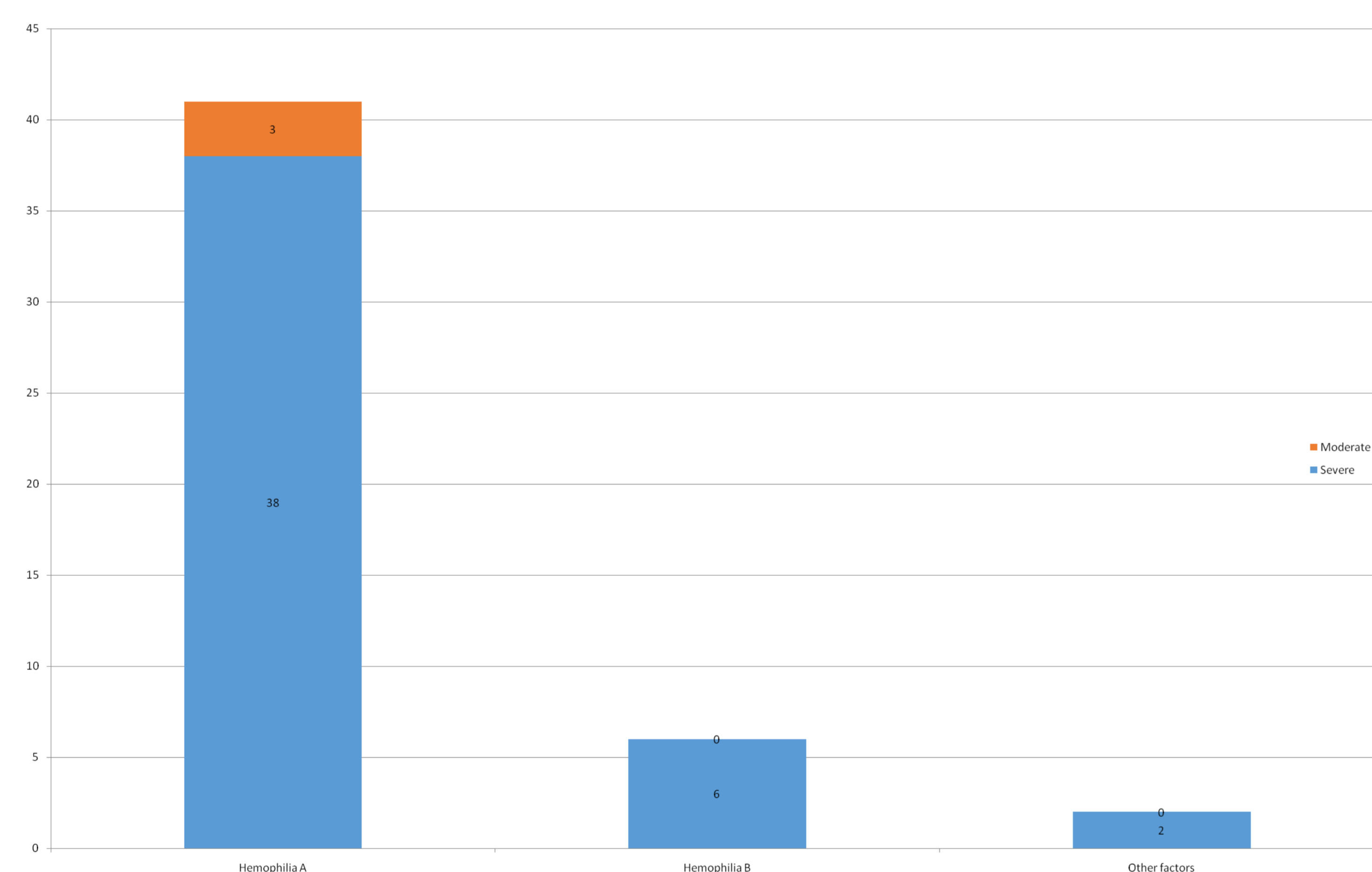
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Introduction and Objectives: Pseudotumours are rare musculoskeletal complications in haemophilia. We present a series of 49 patients with limb or girdle pseudotumours who underwent surgical management at our centre between 1999 and 2014.

Patients and Methods: Clinical data, extent of pseudotumour, surgical details, perioperative factor replacement, and outcome were collected from patient records. Radiological investigations and clinical photos were also scrutinized.

Results:

- 41 patients had hemophilia A (38 severe and 3 moderate), 6 had hemophilia B (all severe), and 2 had other factor deficiencies.
- 12 patients had inhibitors.
- A majority of the patients had involvement of the pelvis and lower limb (Femur/thigh segment- 26, calf/leg segment -9, foot 7 and pelvis-3).
- 5 patients required unexpected additional surgical procedures.



Average perioperative factor requirement

Factor VIII	490 IU/kg	284- 1360
Factor IX	445 IU/Kg	191-710

- Limb ablation was more likely with:**
- Skin breakdown (ulceration/fungation),
- Pathological fracture, extensive skeletal involvement not amenable for reconstruction,
- Distal neurovascular involvement.
- High Inhibitor titre.

Conclusions:

- This case series of hemophilic pseudotumours would constitute one of the largest in recent years .
- Pseudotumours predominantly involve the lower extremity.
- Early diagnosis and intervention may prevent complications that could lead to loss of limb.
- Extensive skeletal destruction and distal neurovascular compromise were likely to result in limb ablation.
- Limb salvage surgery requires adequate resources for prolonged management and a perseverant team.

