UK long bone fracture standards for Duchenne muscular dystrophy (DMD)

formally endorsed by the British Society for Children's Orthopaedic Surgery

Introduction: Duchenne Muscular Dystrophy (DMD) is a complex multisystem disease carrying an increased fracture risk. The principles of fracture management are in line with those already established for frail patients with fragility fractures. Key considerations are: multidisciplinary working; adequate analgesia; fracture management geared to enabling very early return to baseline function; a documented rehabilitation plan. Possible comorbidities including adrenal insufficiency, cardiomyopathy and respiratory impairment should be considered. The priorities in management goals change depending on pre-injury functional level. Pre-existing weight-bearing function should be preserved. Pain-free seating and upper-limb function are the priorities in non-ambulant patients who do not undertake functional standing or assisted transfers.

Inclusions: All patients with DMD presenting to hospital with an acute long-bone fracture.

Standards of Care

1. **All patients must receive effective analgesia.** Regional nerve blockade initiated in the Emergency Department should be considered for lower limb fractures. There is no contraindication to intravenous opioid analgesia.
2. **All patients on long term corticosteroid treatment must be assumed to have adrenocortical suppression**. For these patients, extra doses of corticosteroids must be given on presentation with a fracture to cover the stress response. Further intravenous doses must be given during any operation.
3. **Specialist advice on medical and surgical management should be sought urgently** by contacting the Muscle Team which routinely cares for the patient and the regional specialist children's orthopaedic team to advise on fracture management.
4. **There should be a documented rehabilitation plan** covering early mobilisation to community care. The plan should be included in discharge documentation and copied to the patient/ family.
5. **The aim of fracture management** is to enable pre-injury weight-bearing, seating and upper limb function within 36 hours of admission. (cf fragility fracture BOAST)
6. **Cast treatment** for stable fractures in functional alignment should be conducted with lightweight circumferential casts within 36 hours of admission with joints immobilised in the best achievable functional position.
7. **Operative stabilisation** should be considered for long bone and periarticular fractures which cannot be stabilised by non-operative means, following discussion of the relative risks with specialist clinicians, patient and family.
8. **Suxamethonium must not be given and volatile anaesthetic agents should be avoided in DMD.** Total intravenous anaesthesia (TIVA) and regional anaesthetic blocks are recommended.
9. **Surgical management** should aim to enable early pre-injury level weight-bearing, seating and upper limb function (see standard 5)
10. **Operative stabilisation** should follow the established principles for fragility fracture management (protect the full length of the bone, combined nail/plate fixation or double plating etc.)
11. **Fat embolism** occurs more frequently in patients with DMD and the signs may be subtle. The risk and the plan to manage the condition if it occurs should be documented in the patient record.

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