363.00

ALPHA-MANNOSIDOSIS, DELAYED BONE HEALING AND A NEW VARIANT OF DYSTOSIS MULTIPLEX IN TWO BROTHERS

R.O. Sundaram, V. Bhalaik, H.P.J. Walsh

Department of Orthopaedics, Toyal Liverpool Children's Hospital Liverpool, United Kingdom
rathers74@yahoo.co.uk

Alpha-Mannosidosis is a rare autosomal-recessive condition described first in 1967. There have been more that 50 reported cases of orthopaedic manifestations commonly known as 'dystosis multiplex'. We describe complications following a varus de-rotational femoral osteotomy, and new variant of dystosis multiplex; in two brothers with Alpha-mannosidosis. Case 1: A 14 year old boy diagnosed with Alpha-mannosidosis presented with right hip pain. Radiographs of the hips showed bilateral hip dysplasia, the right worse than the left, with uncovering of the acetabulum over the right femoral head. The patient proceeded to have a right varus de-rotational femoral osteotomy. The patient suffered complications of wound break down and formation of sterile cysts. The femoral osteotomy failed to unite after 6 months; 2 injections of bone marrow were undertaken at the osteotomy site, which were unsuccessful. After immobilization in a hip spica for 6 weeks; union was eventually achieved at 1 year. We demonstrate that delayed union may be a feature in patients with Alpha-mannosidosis. 6 years post surgery the patient's hip symptoms are mild and has a good range of hip movement. Case 2: The 13 year old brother of case 1 presented with back pain, flat feet and bilateral foot pain. Radiographs showed marked osteochondral changes of the lumbar and cervical spine, the right foot showed evidence of avascular necrosis of the navicular. The patient had been managed conservatively; the right foot with modified foot wear. Back and feet symptoms are mild 5 years after initial diagnosis.