Management of Osteofibrous Dysplasia – determining current practice trends in Europe and establishing a multinational disease registry

Background

Osteofibrous dysplasia (OFD) is a rare, benign fibro-osseous lesion, accounting for about 0.2% of all primary bone tumours. The lesions appears to almost exclusively affect the tibial diaphysis, with occasional involvement of the fibula, and only a few reports of other bones being affected. The condition usually presents with swelling or anterior bowing of the leg, which may be associated with pain.

Adamantinoma (AD) is a rare low-grade malignant primary bone tumour (<1% of all primary bone malignancies), usually occurring in the second and third decades of life. The predilection for the anterior tibial diaphysis is similar to OFD, with over 80% of cases involving this location. The presenting clinical features of pain, swelling and deformity are also similar to those of OFD.

The relationship between OFD and AD remains controversial. The similarities in their location, imaging characteristics and histology have led many to suggest that the two are related, and are entities on a spectrum of the same disease. The formulation of the intermediary OFD/LA sub-type was felt by many to prove the existence of this spectrum, with progression from OFD to AD.

Based upon this risk of possible transformation, some surgeons advocate radical extraperiosteal resection for OFD, necessitating complex reconstruction for the resultant defect, either with distraction osteogenesis, endoprostheses or massive bone grafts. Others, including Campanacci, who first reported OFD in detail, have advocated conservative management of these lesions, with surgical treatment, such as subperiosteal resection or curettage, reserved for symptomatic lesions only.

From the patient's perspective, this lack of consistency amongst the medical profession would undoubtedly be a source of angst and confusion; how could one surgeon recommend massive surgery for a condition, whilst another argues that it should be left alone, and who should they believe?

Study Objective & Method

The purpose of this study is to determine variation in the management of OFD across specialist centres in Europe, and to quantify its extent. We will do this by collating data about surgeons' current practices in the management of OFD, looking specifically at referral pathways, diagnostic work-up, surgical management and follow-up protocols. This will allow identification of areas of consensus and controversy.

This information will then form the basis for further study to improve the management of the child with OFD, in an attempt to eliminate the controversy and reduce the dramatic variations in practice to ensure all patients receive the most appropriate care.

The tailored design method will be used for survey design and distribution. The survey distributed to all members of EPOS. Members of the European Musculo-Skeletal Oncology Society will also be invited to participate in the study.

The planned timetable for the study is:

April 2014	Field testing of survey
June 2014	General distribution of survey
April 2015	Presentation of results of the survey