The importance of patient reported functional outcome in paediatric desmoid fibromatosis

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Dear Editor,

Desmoid fibromatosis (DF) is a heterogeneous condition with lack of consensus regarding therapy. Fatality is rare but repeated surgery prevalent. Functional assessment for this group is not well established, although we believe it to be an important marker of treatment success. At present there is no validated objective or subjective functional assessment tool designed for benign but locally invasive disease.

We reviewed self-assessed functional outcome using the Toronto Extremity Salvage Score (TESS) in 19 young people (<21 years, treated 2003-2010), with a histological diagnosis of DF in an extremity. Each of the 30 questions within the TESS is a measure of the difficulty that an individual has performing a task.[1] Scores per question range from a minimum of 1 to a maximum of 5.

Median time from diagnosis to TESS was 97 months (interquartile range 26-143 months). TESS was lower in children who underwent surgical resection compared with those who did not. Children with progressive disease, 5 or more treatment modalities, or greater than 6 events (progressive disease) had lowest outcome scores. TESS of those children receiving cytotoxic and non-cytotoxic therapies was high (median TESS 75.5%), despite a trend towards advanced inoperable disease. TESS was lower in those receiving radiotherapy.

Within orthopaedics, patient reported outcome measures are becoming increasingly important; driven by the Department of Health they are also being utilised to direct finite resources. Given that this disease is very rarely fatal, with so many different treatment options, we believe that a greater knowledge of functional assessment is an important step to improving outcome. Longitudinal data may aid optimisation, validation and development of treatment strategies. Longitudinal functional assessment should not be time-consuming, with completion of TESS designed to take under 10 minutes. To facilitate ease of completion, we propose a touch screen electronic tablet interface for patient input of questionnaires while waiting for outpatient clinical review. We suggest an annual formal objective assessment of function with additional assessment prior to a new therapeutic modality.

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