

Management and surveillance of abdominal, retroperitoneal and pelvic schwannomas: an international cohort study from the Trans-Atlantic Australasian Retroperitoneal Sarcoma Working Group (TARPSWG)

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Aim

- ▶ Determine a consistent international management policy for abdominal, retroperitoneal and pelvic schwannomas
- ▶ Differences in specialist centre management created an opportunity
 - ▶ To study early surgery and the value of post-operative surveillance
 - ▶ And conversely, radiological monitoring / growth characteristics



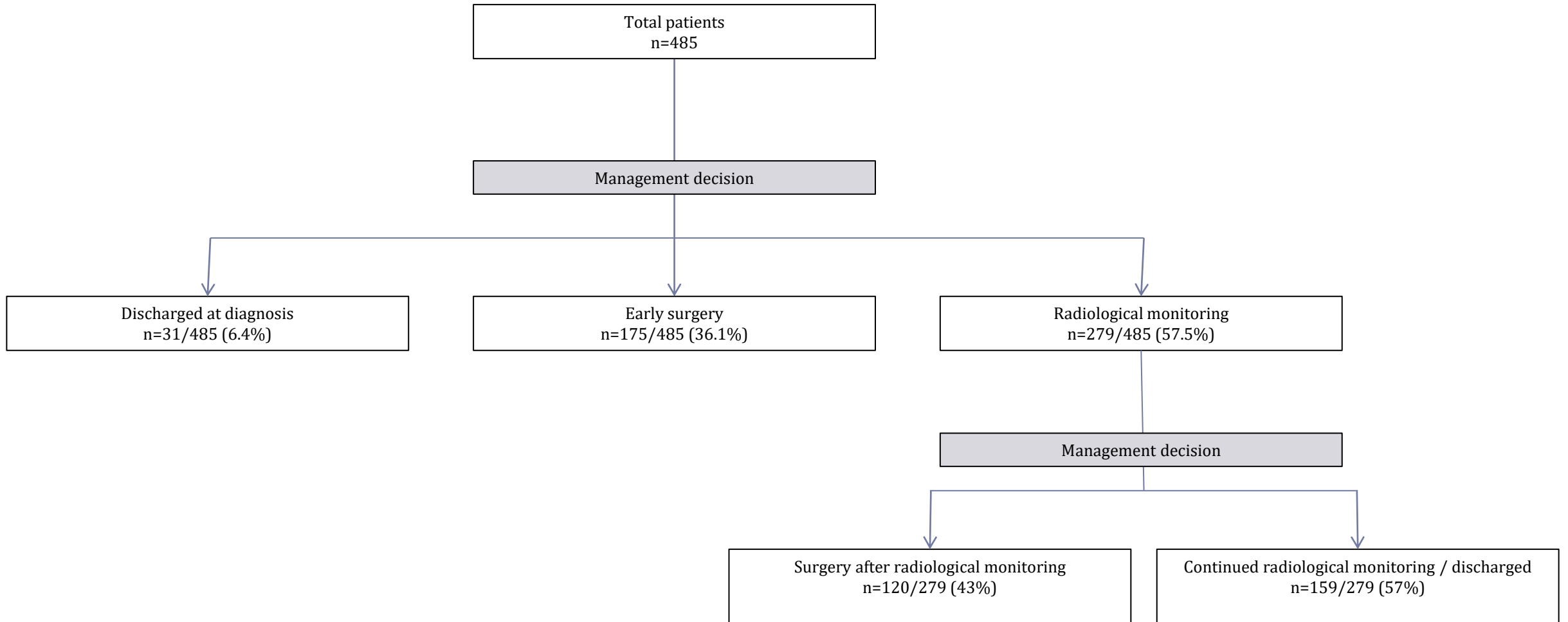
Contributing centres – thank you!

Name of Centre	Number of Patients
Brigham and Women's Hospital/Dana-Farber Cancer Institute. Boston, USA	26
Emory University Hospital. Atlanta, USA	83
Institute of Oncology Ljubljana. Ljubljana, Slovenia	21
Istituto Nazionale dei Tumori. Milan, Italy	37
Mayo Clinic. Jacksonville, USA	23
Moffitt Cancer Centre. Tampa, USA	13
Mount Sinai Hospital. Toronto, Canada	63
Netherlands Cancer Institute. Amsterdam, The Netherlands	10
Ottawa Hospital Research Institute. Ottawa, Canada	55
Peter MacCallum Cancer Centre. Melbourne, Australia	17
Queen Elizabeth Hospital. Birmingham, UK	52
Royal Marsden Hospital. London, UK	85

Total 485 from 12 centres



Flow diagram of patient management



Longitudinal schwannoma growth

236 patients (904 scans)

Tumour volume increased significantly over time ($p < 0.001$)

Median of 10.5% p.a. (95% CI: 9.4% - 11.6%)

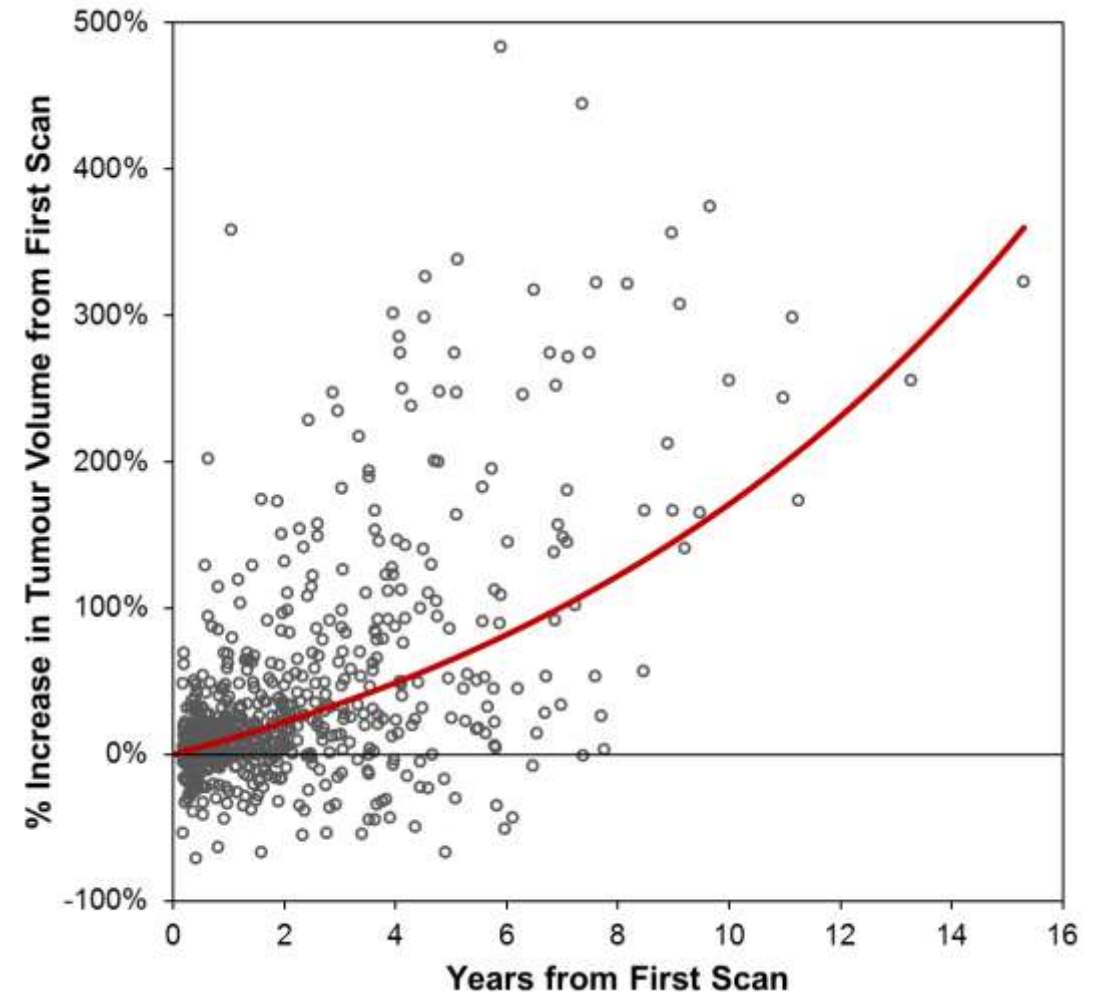
146 patients had at least 3 scans (median growth 10% p.a.)

17% declined in volume

12% stable +/- 2%

47% variable growth gradient of 3-20% p.a.

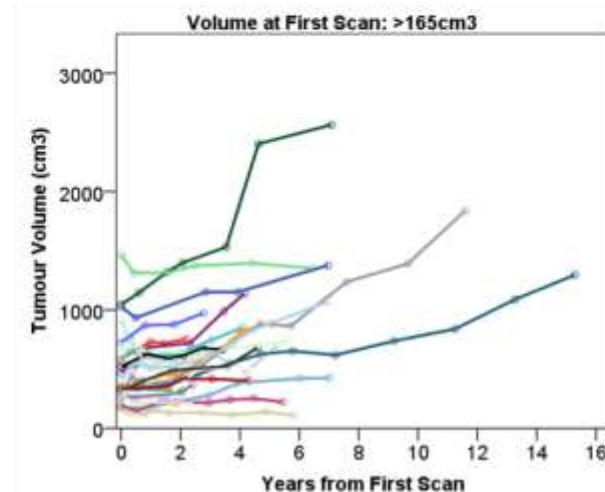
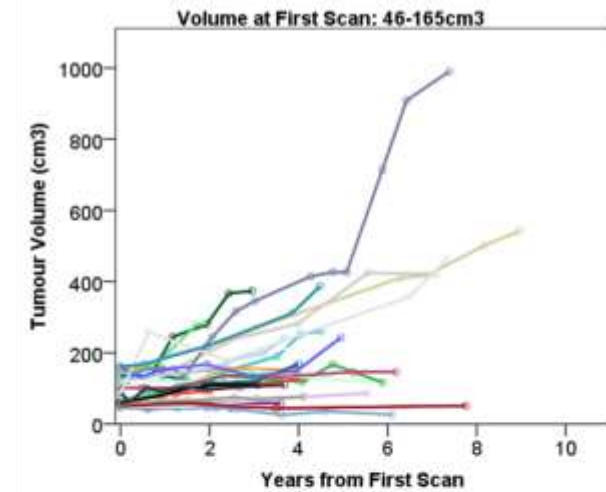
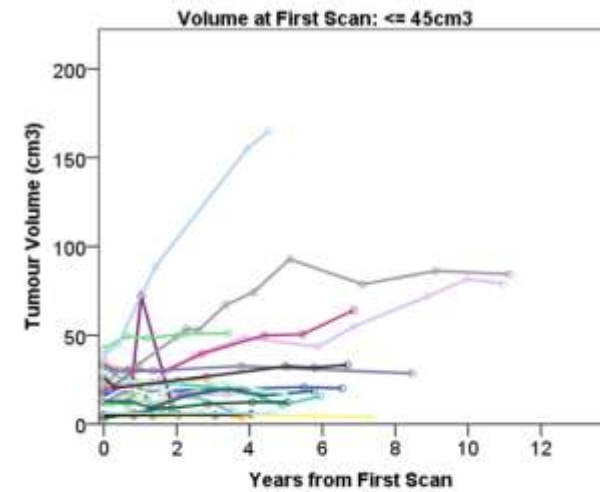
23% rapid growth of >20% p.a.



Marked individual variability

Patients with five or more scans based on initial tumour volume

No predictors of growth at presentation

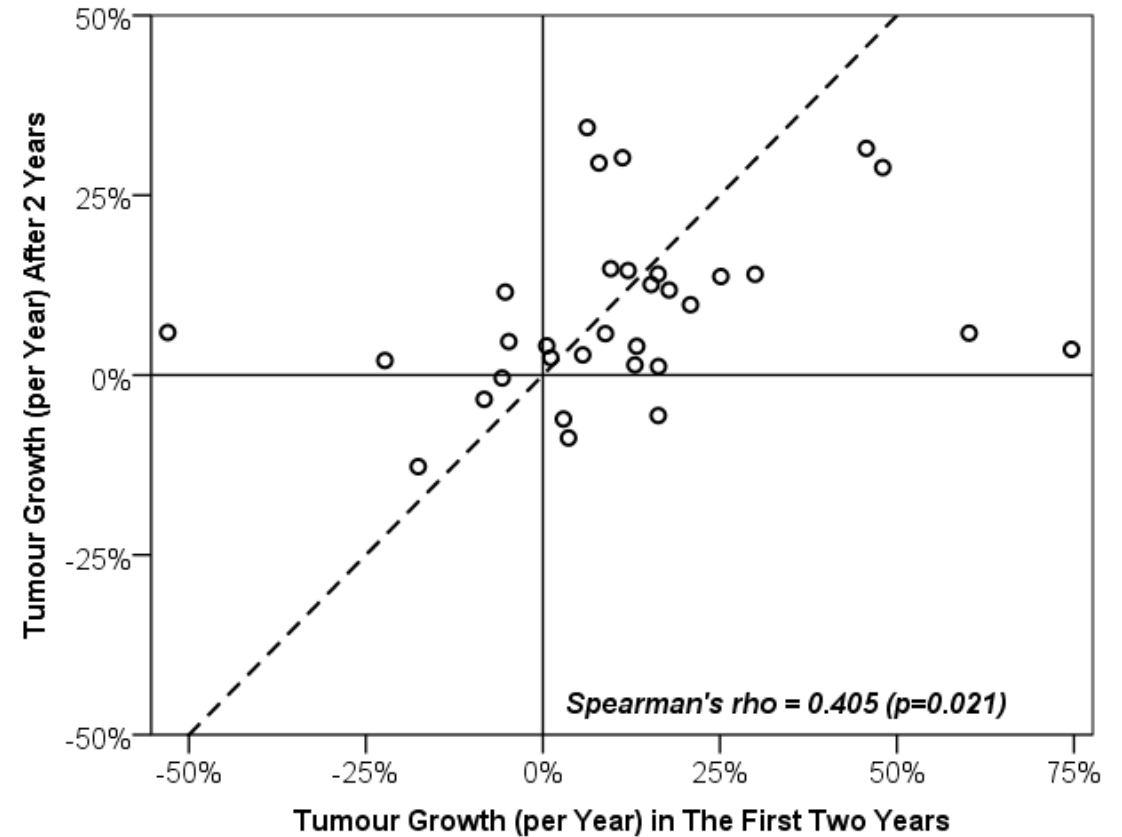


Using imaging to predict future growth

Comparisons of short and long term growth

Growth rate over two years (based on a series of three scans) significantly correlates with subsequent expansion

No significant difference between the short and long-term growth rates $p=0.021$



Bland-Altman analysis

Predictors of surgery

- ▶ 61% underwent (36% at presentation and 43% of the monitored cohort)
- ▶ Surgery occurred relatively soon after monitoring (median of 9.7 months)
- ▶ Predictors of surgery
 - ▶ Significantly more likely
 - ▶ Symptomatic (versus co-incidental) HR 1.53 (1.21 - 1.93)
 - ▶ Tumour growth >20% per annum HR 10.2 (1.33 – 77.7)
 - ▶ Significantly less likely
 - ▶ Major nerve involvement HR 0.56 (0.39 - 0.80)
- ▶ Predictors of operative complications
 - ▶ Tumour growth >20%
 - ▶ Originating from major nerve
 - ▶ Major nerve division
 - ▶ Division of major artery – high grade complication



Post operative surveillance

- ▶ R0/R1 resections

- ▶ Accurately predicted at pre-operative imaging
 - ▶ 47% (n=124) resections entered post-operative surveillance imaging
 - ▶ 3% had evidence of residual disease at base-line imaging
 - ▶ 3% (n=4) developed radiological evidence of recurrent disease
 - ▶ Kaplan-Meier estimated long-term recurrence rates of 2% and 7% at 2 and 5 years

- ▶ R2 resections

- ▶ 88% (n=21) entered surveillance 15 had radiological evidence of residual disease
- ▶ No interventions during the study period



Recommendations

- ▶ Early indications for surgery include
 - ▶ Symptomatic tumour at presentation
 - ▶ Diagnostic uncertainty
 - ▶ Existing evidence of rapid expansion
 - ▶ Patient preference after discussing options
- ▶ Where an indication for surgery at presentation exists
 - ▶ Surgery should proceed with adequate patient counselling
 - ▶ Consideration given to a planned R2 resection if major nerves or vessels would otherwise need to be sacrificed.



Recommendations

- ▶ In the absence of an indication for early surgery
 - ▶ Diagnosis of a schwannoma should be confirmed on biopsy
 - ▶ Patient enrolled in to a programme of monitoring imaging for at least 2-years in order to predict individualised tumour growth characteristics
 - ▶ If not operated after 2 years, multi-disciplinary discussion should then occur to consider future resectability and likely induced morbidity of surgery, coupled with patient age, wishes and specific tumour location.
- ▶ Post-operative surveillance imaging for patients with R0/R I resection holds little benefit beyond a baseline scan and should not be routinely recommended
- ▶ Post-operative surveillance imaging after R2 resection should be considered on a pragmatic basis





Mesenteric Sarcoma Project



Rationale

- ▶ Patient characteristics / spectrum of histology and outcomes are largely unknown - although anecdotally poor
- ▶ Extremely rare – will require a large number of collaborating centres
- ▶ Inclusion criteria
 - ▶ All primary mesenteric sarcomas that are clearly arising from the mesentery and those where the origin has a degree of uncertainty i.e. abutting the bowel
 - ▶ All sarcoma pathology apart from GISTs
- ▶
- ▶ Exclusion criteria
 - ▶ Tumours arising from the bowel wall that have invaded the mesentery
 - ▶ Patients under 18 years old on the time of diagnosis
 - ▶ GISTs and fibromatosis



Data so far – plenty of time to participate!

17 centres have indicated interest

Data from 8 centres

31 cases in total – 3-4 per centre

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Primary mesenteric sarcomas		
Centre	Name	Data received?
Birmingham, UK	Samuel Ford/Hannah Tattersall	
LA, USA	William Tseng	Y
Milan, Italy	Alessandro Gronchi	Y
Ottawa, Canada	Caroline Nessim	
Venice, Italy	Marco Rastrelli	
Napoli, Italy	Massimiliano Di Marzo	
Victoria, Australia	David Gyorki	
Torino, Italy	Giovanni Grignani	
Barcelona, Spain	Jose Antonio Gonzalez	
Peking, China	Cheng-Hua Luo	
Miami, USA	Neha Goel	
Amsterdam, Netherlands	Winan van Houdt	Y
Georgia, USA	Kenneth Cardona	Y
Leiden, Netherlands	Yvonne Schrage	Y
Korea	SoHee Lim	
Florida, USA	Sanjay Bagaria	Y
Ljubljana, Slovenia	Marko Novak	Y