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## INTRODUCTION

- Desmoids are locally aggressive tumors derived from fibroblast cells
- Desmoids can arise in the abdomen and extremities
- In the extremities and abdominal wall, it has been shown that 25-30% of desmoids regress, 10-15% rapidly progress and the majority remain stable
- Across all anatomical locations it is suggested that 46% of desmoid undergoing active surveillance eventually require treatment
- Currently, there are no high-power studies reporting the natural course and outcomes of mesenteric and intra-abdominal desmoids

## METHODS

Date was collected from TARPSWG Sarcoma Centres from Jan 2008 to June 2020 (n=762)

- Exclude (n=120):
- Non-desmoid tumours
  - Incorrect primary site (extremity, abdominal wall)
  - No scan dimensions for OBS/MED patients
  - Missing surgery date, margins

Cases Included (n=642)

- Cohort classification
- Classified upon initial collection.
  - If >180 days from First Scan to SURG, case reclassified to OBS.

OBS (n=153)	MED (n=144)	SURG (n=345)
OUTCOMES		
1. Desmoid status from first to last scan (RECIST V1.1)	1. Desmoid status from first to last scan (RECIST V1.1)	1. Progression-free survival (PFS), with progression on R2 being an event of recurrence.
2. Crude Cumulative Incidence of Surgery (CCI) analysis	2. Progression-free survival (PFS) analysis	
3. Progression-free survival (PFS) analysis		

### Statistical Analysis

- KM curves, multivariable Cox models (PFS), crude cumulative incidence and Fine & Gray models (CCI of Surgery) were performed across initial management cohort to assess the association of age, sex, FAP status, primary site, medical treatment (MED only), and completeness of resection (SURG only) with the indicated endpoints.

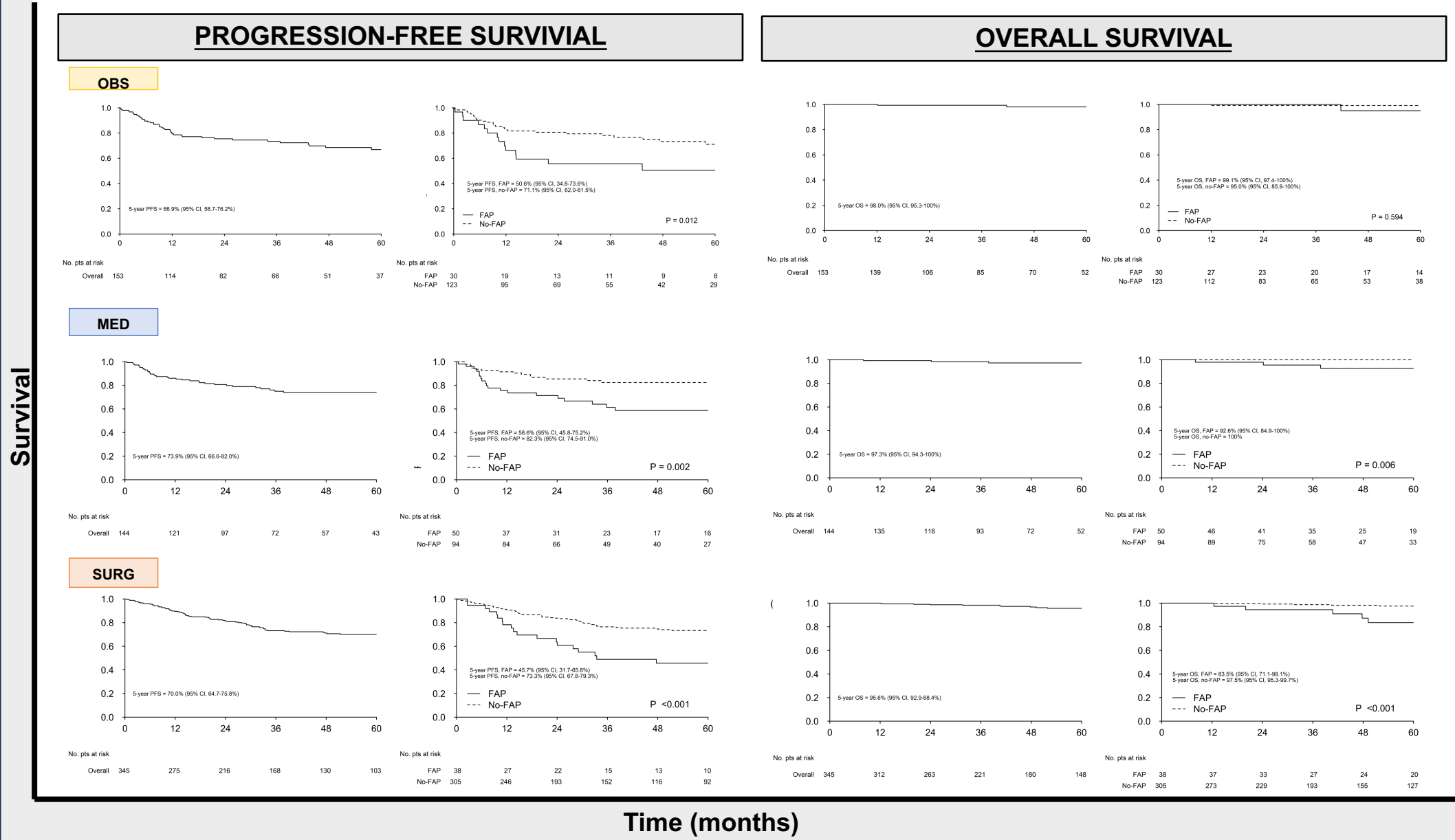
## BASELINE CHARACTERISTICS

**Table 1.** Baseline Demographic, Clinical, and Pathological Characteristics (n=642)

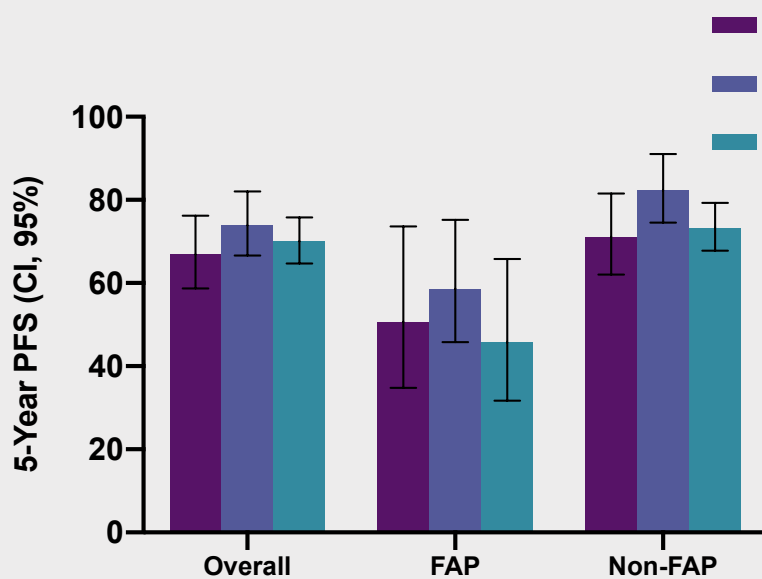
Characteristic	Result
<b>Sex, No. (%)</b>	
Male	335 (52.2)
Female	307 (47.8)
<b>Age, median (IQR), years</b>	
	42.0 (31.0-55.0)
<b>Follow-up Period, median (IQR), months</b>	
All Cohorts	50.0 (25.3-87.0)
OBS	44.8 (21.6-81.4)
MED	50.7 (27.8-87.2)
SURG	52.8 (27.2-91.1)
<b>Initial Management Cohort, No. (%)</b>	
OBS	153 (23.8)
Observation only	77 (50.3)
Observation then medical management	30 (19.6)
Observation then surgery	32 (20.9)
Observation then medical + surgery	14 (9.2)
MED	144 (22.4)
Medical only	108 (75.0)
Medical then surgery	36 (25.0)
SURG	345 (53.7)
SURG (R0-R1)	278 (80.6)
Elective	229 (82.4)
Emergency	49 (17.6)
SURG (R2)	67 (19.4)
Elective	51 (76.1)
Emergency	14 (20.9)
Not indicated	2 (3.0)
<b>Surgery Summary Over Total Course, No. (%)</b>	
Complete Resection (R0-R1)	336 (52.3)
Incomplete Resection (R2)	91 (14.2)
No surgery throughout course	215 (33.5)
<b>FAP, No. (%)</b>	
Yes	118 (18.4)
No	522 (81.3)
Unknown	2 (0.3)
<b>Desmoid Site, No. (%)</b>	
Mesenteric	416 (64.8)
Retroperitoneal	98 (15.3)
Pelvic	78 (12.1)
Intra-peritoneal	46 (7.2)
Not available	4 (0.6)

## RESULTS

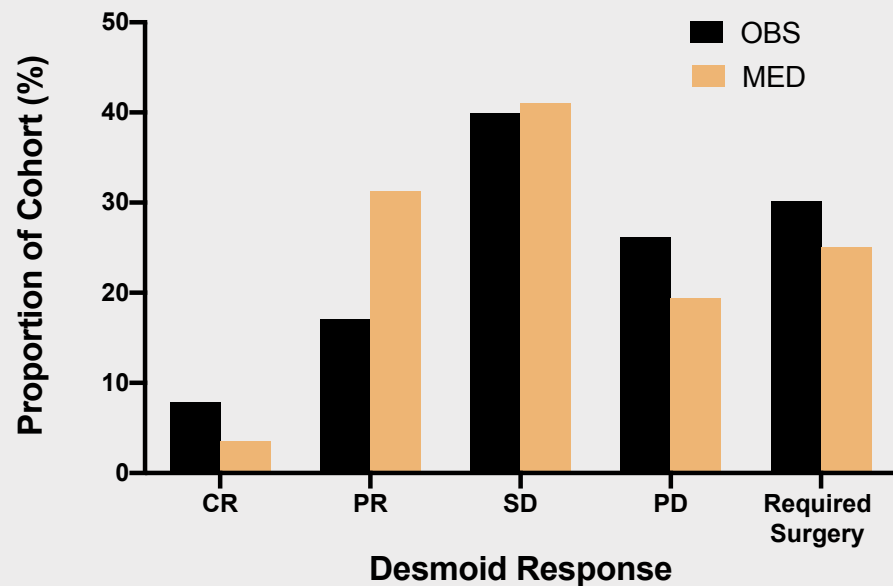
**Figure 1.** Overall and Progression-Free Survival by Cohort and FAP Status



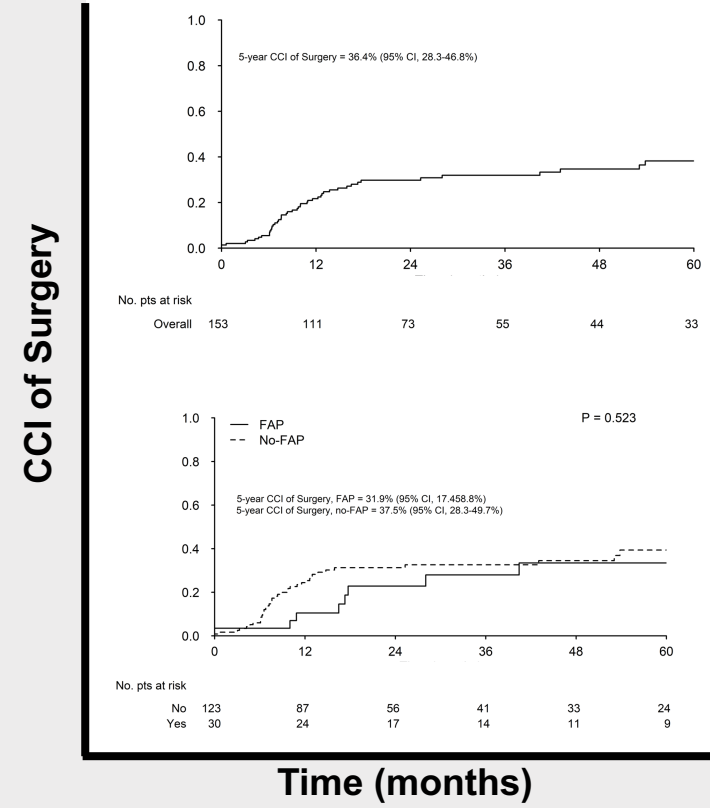
**Figure 3.** 5-Year PFS by Cohort and FAP-Status



**Figure 4.** Response During Non-Surgical Management



**Figure 5.** Crude Cumulative Incidence of Surgery of OBS Cohort

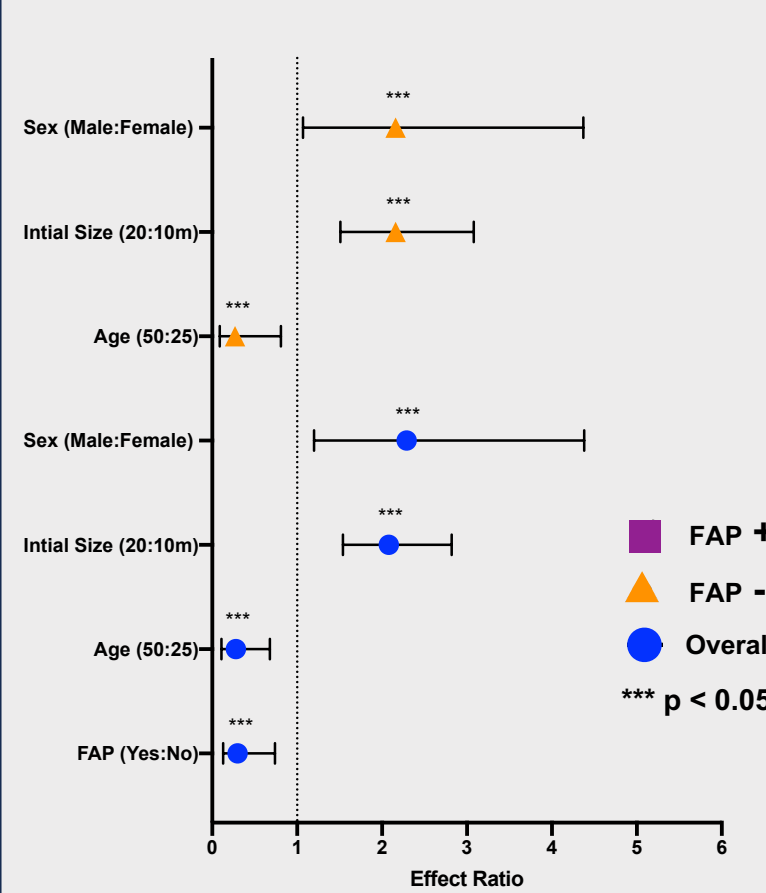


Across all cohorts, FAP+ patients had a lower PFS than non-FAP patients. 5-year PFS was highest in MED, but with values falling within each others 95% CI.

OBS cohort shows similar rates of partial regression (PR) and disease stabilization (SD) to desmoids of the extremity and abdominal wall.

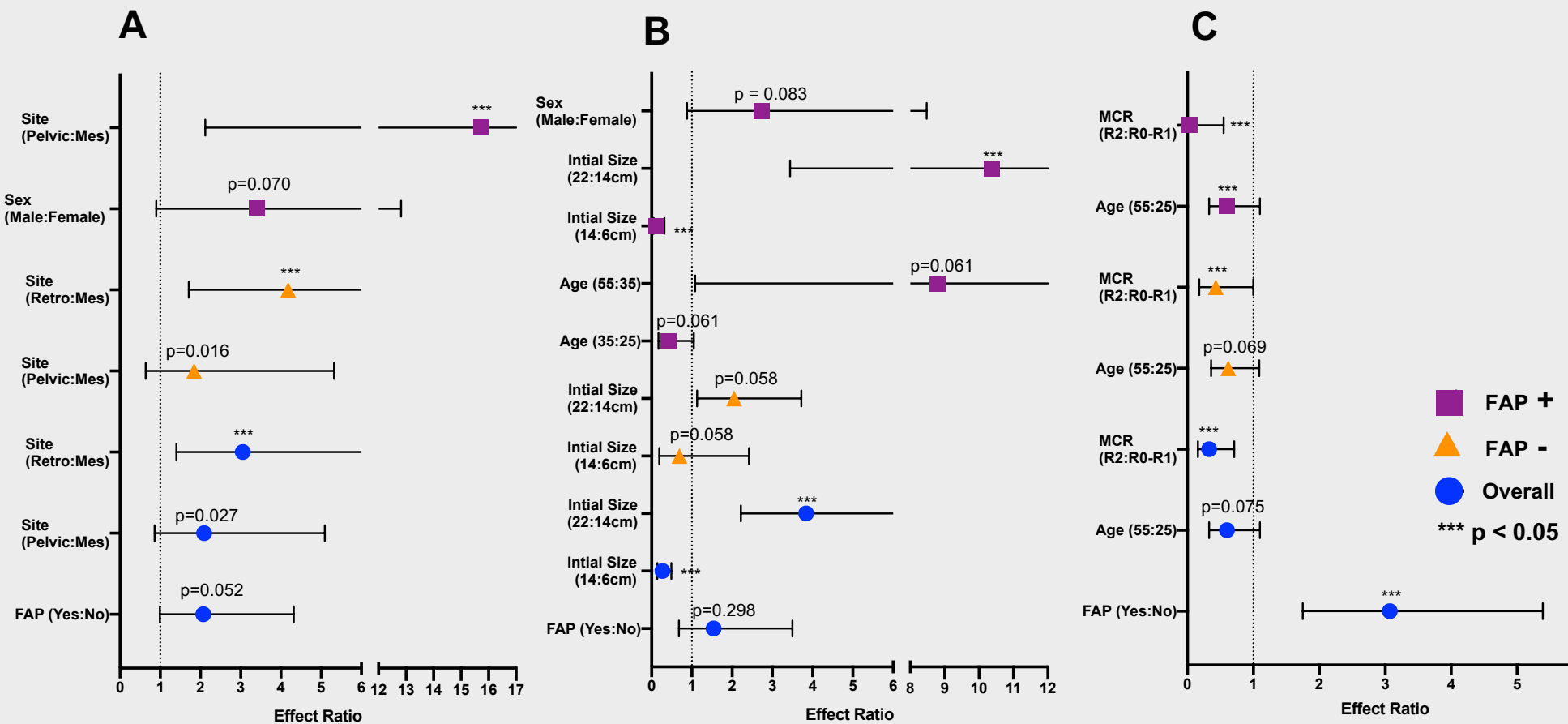
In the OBS group the incidence of surgery is about 36%, with a slightly higher CCI in FAP patients.

**Figure 6.** Main Prognostic Events on Time to Surgery of OBS



Overall, FAP -, older age, smaller initial size and male sex were associated to a lesser risk to undergo to surgical resection.

**Figure 7(A-C).** Main Prognostic Events on PFS of the OBS (A), MED (B) and SURG (C) Cohorts and FAP Status



Across all cohorts FAP had an adverse prognostic effect with significance in the SURG cohort, and nearing significance in the OBS cohort (p=0.052). In the OBS cohort, site was a major significant prognostic factor with both retroperitoneal and pelvic having worse outcomes than mesenteric. Initial size shown a "U" shaped relationship with PFS in MED, with increasing protective effect from 8 to 14 cm, and adverse thereafter. Lastly, completeness of surgical resection (R0/R1) was not associated with more favorable outcome.

## CONCLUSIONS

- Active surveillance in intra-abdominal desmoids is feasible and yields similar percentages of spontaneous disease stabilization and regression as for desmoid tumors at other anatomical sites
- Observed deaths on overall survival analysis mostly occurred in FAP+ patients, with FAP+ patients also having a lower 5-Year PFS than FAP- patients across all cohorts
- FAP was identified as and adverse event in across all initial management cohorts
- 5-year PFS is similar across all initial management cohorts, suggesting that they are all viable treatment options
- In the OBS cohort, site was a major significant prognostic factor, with both retroperitoneal (especially in no-FAP) and pelvic (especially in FAP) having worse outcomes than mesenteric
- Surgery is option as initial management when surgical morbidity is acceptable in patients affected by sporadic desmoids, especially if located to the mesentery or retroperitoneum.

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