

Tumeurs Desmoides et Grossesse

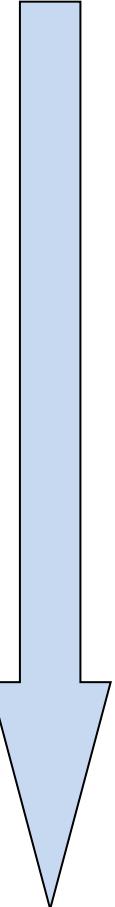
Dimitri Tzanis

Sarcomes et Tumeurs Complexes



INDICATIONS OF SURGERY CHANGED OVER TIME

2005

- 
1. Surgery when feasible
 2. Wait and see for recurrent but stable lesion
 3. Wait and see for primary irresectable lesion
 4. The effect of surgical margins is unclear and a conservative approach is preferable
 5. Wait and see for selected primary resectable lesions
 6. **Surgery is no more « standard treatment »**

Bonvalot EJSO 2008, Fiore ASO 2009, Salas JCO 2012, ESMO Guidelines 2014,
EORTC guidelines 2014 (EJC) and update 2017 (Ann Oncol)

2017

Extra-abdominal primary fibromatosis: Aggressive management could be avoided in a subgroup of patients[☆]

S. Bonvalot ^{a,*}, H. Eldweny ^a, V. Haddad ^b, F. Rimareix ^a, G. Missenard ^a, O. Oberlin ^c, D. Vanel ^d, P. Terrier ^e, J.Y. Blay ^f, A. Le Cesne ^g, C. Le Péchoux ^h

76 pts: Surgery

13 pts: Surgery + RT

23 pts: NS (ST or W&S)

3 yrs EFS: R0:65% NS:68%

Growth arrest: 2/3 NS

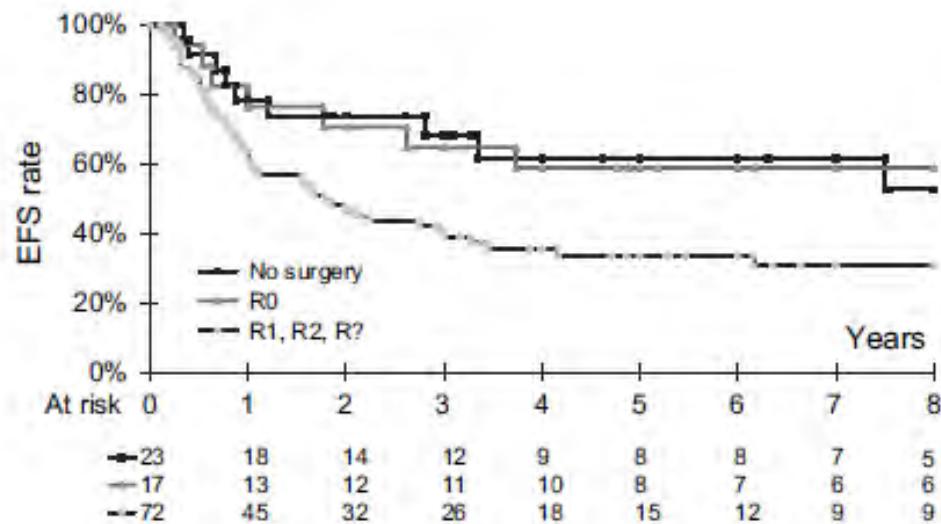


Figure 2. Event-free survival according to the quality of surgery (R0 versus no-surgery versus R1, R2, R not evaluated) (R? = R not evaluated).

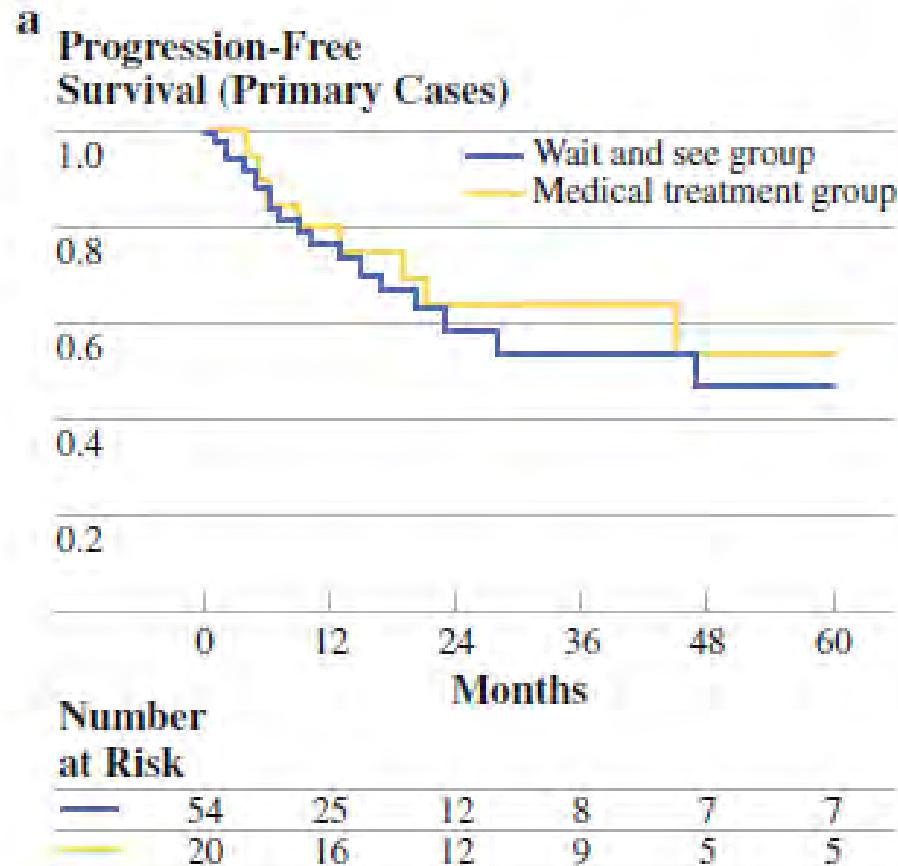
ORIGINAL ARTICLE – BONE AND SOFT TISSUE SARCOMAS

Desmoid-Type Fibromatosis: A Front-Line Conservative Approach to Select Patients for Surgical Treatment

Marco Fiore, MD¹, Françoise Rimareix, MD², Luigi Mariani, MD³, Julien Domont, MD⁴, Paola Collini, MD⁵, Cecile Le Péchoux, MD⁶, Paolo G. Casali, MD⁷, Axel Le Cesne, MD⁴, Alessandro Gronchi, MD¹, and Sylvie Bonvalot, MD, PhD²

142 pts (74:PD, 68:RD)
W&S: 83 pts MT:59 pts

- 5-year PFS: 49.9% for the W&S group (these pts were over treated before)
- 5-year PFS: 58.6% for the medical treatment (MT) group
- **2/3 pts with primary avoid any surgery**
- **50 % pts with primary avoid any treatment**
- For pts who progressed, median TTP: 14 months





Original Research

Surgical versus non-surgical approach in primary desmoid-type fibromatosis patients: A nationwide prospective cohort from the French Sarcoma Group

Nicolas Penel ^{a,*}, Axel Le Cesne ^b, Sylvie Bonvalot ^c, Antoine Giraud ^d,
Emmanuelle Bompas ^e, Maria Rios ^f, Sébastien Salas ^g, Nicolas Isambert ^h,
Pascaline Boudou-Rouquette ⁱ, Charles Honore ^b, Antoine Italiano ^j,
Isabelle Ray-Coquard ^k, Sophie Piperno-Neumann ^l, François Gouin ^{m,n},
François Bertucci ^o, Thomas Ryckewaert ^a, Jean-Emmanuel Kurtz ^p,
Françoise Ducimetiere ^k, Jean-Michel Coindre ^q, Jean-Yves Blay ^k



771 patients
Surgery 359
Wait and see 388

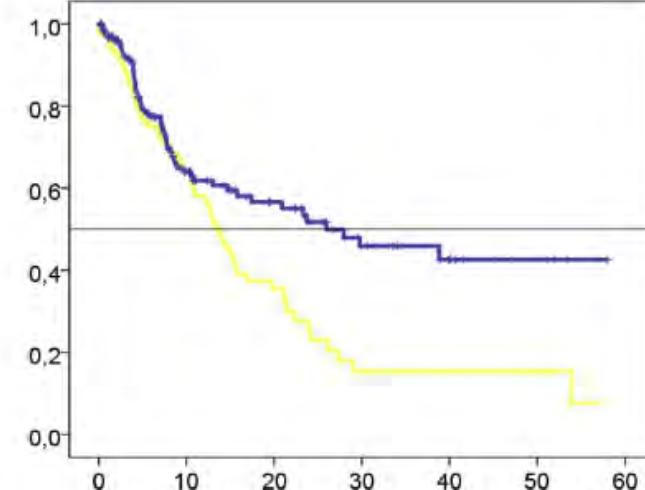


Fig. 1. Outcome of patients with unfavourable locations. Yellow line represents surgery as first treatment and blue line represents initial wait-and-see approach. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

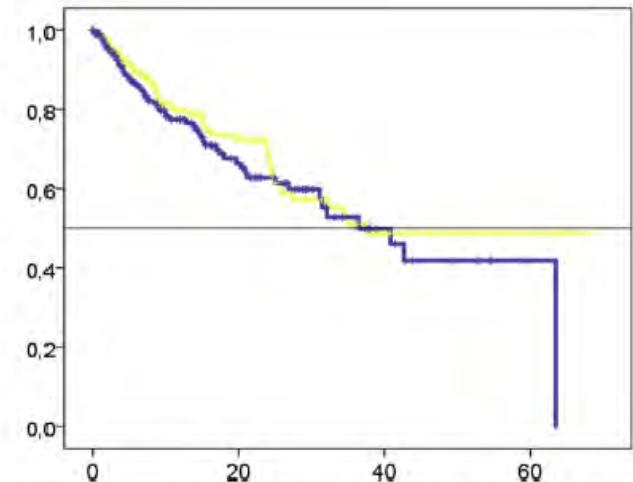
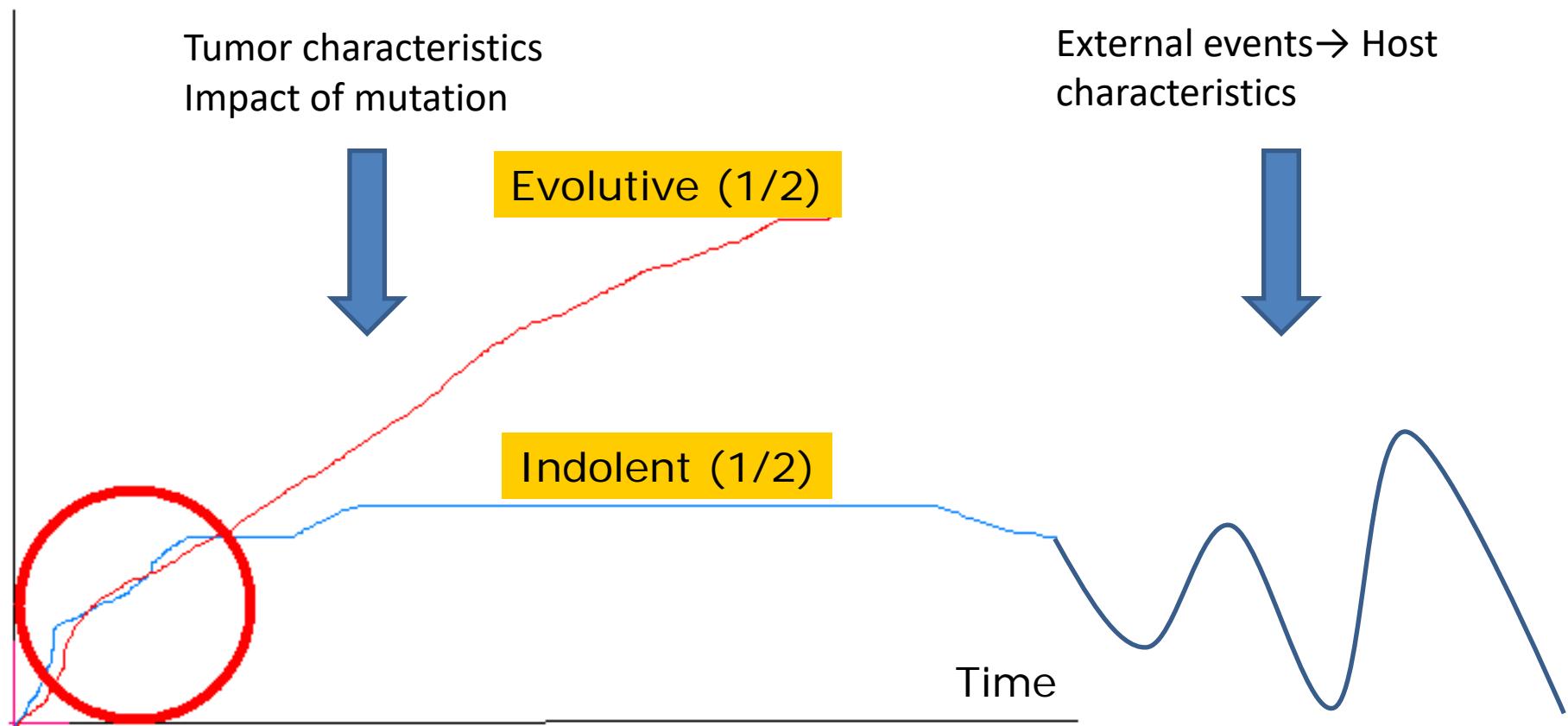


Fig. 2. Outcome of patients with favourable locations. Yellow line represents surgery as first treatment and blue line represents the initial wait-and-see approach. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

Evolution

Tumor size



Prognostic Factors Influencing Progression-Free Survival Determined From a Series of Sporadic Desmoid Tumors: A Wait-and-See Policy According to Tumor Presentation

Sébastien Salas, Armelle Dufresne, Binh Bui, Jean-Yves Blay, Philippe Terrier, Dominique Ranchere-Vince, Sylvie Bonvalot, Eberhard Stoeckle, Louis Guillou, Axel Le Cesne, Odile Oberlin, Véronique Brouste, and Jean-Michel Coindre

Table 4. Multivariate Progression-Free Survival Analysis

Variable	Crude HR	95% CI	P
Median age	1.97	1.36 to 2.84	<.001
Median size	1.64	1.13 to 2.36	.008
Tumor site			
Abdominal wall			
Intra-abdominal tumor	1.95	0.92 to 4.15	.084*
Extra-abdominal tumor	2.55	1.48 to 4.4	<.001

Abbreviation: HR, hazard ratio.

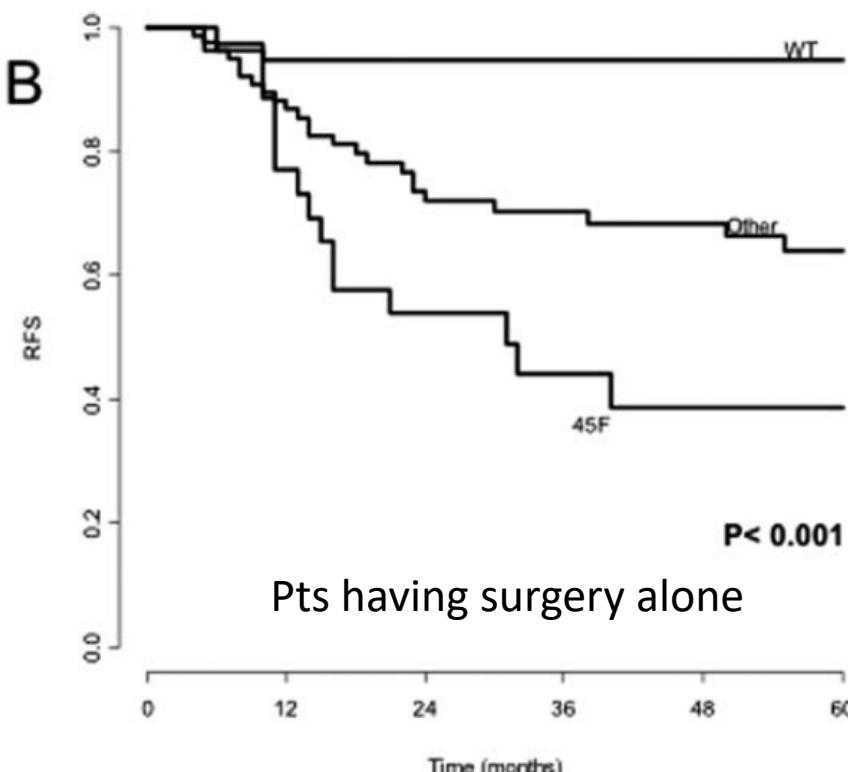
*Not significant.

CTNNB1 45F Mutation Is a Molecular Prognosticator of Increased Postoperative Primary Desmoid Tumor Recurrence

An Independent, Multicenter Validation Study

Chiara Colombo, MD¹; Rosalba Miceli, PhD²; Alexander J. Lazar, MD, PhD^{3,4}; Federica Perrone, PhD⁵; Raphael E. Pollock, MD, PhD^{4,6}; Axel Le Cesne, MD⁷; Henk H. Hartgrink, MD⁸; Anne-Marie Cleton-Jansen, PhD⁹; Julien Domont, MD⁷; Judith V. M. G. Bovée, MD⁹; Sylvie Bonvalot, MD¹⁰; Dina Lev, MD^{4,11}; and Alessandro Gronchi, MD¹

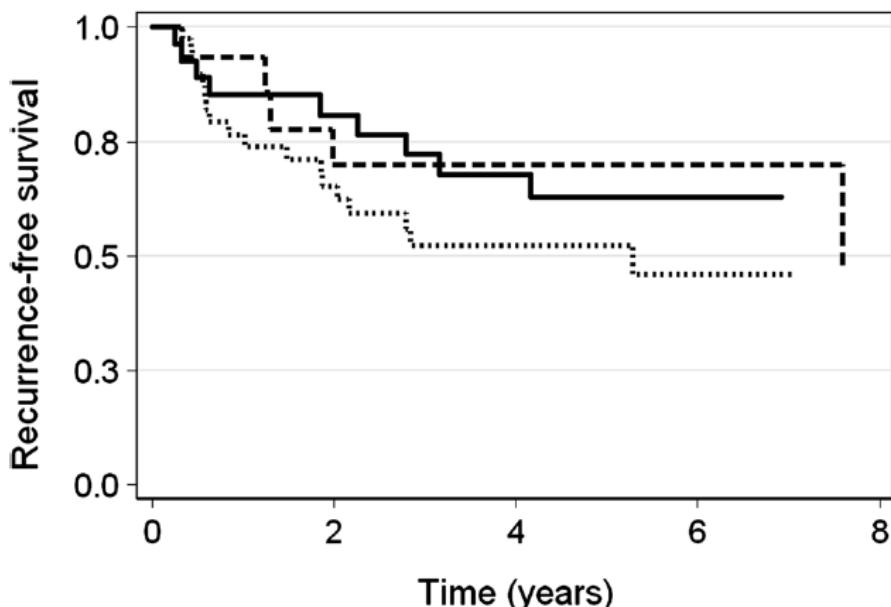
- T41A
- S45F
- S45P



ORIGINAL ARTICLE

Pregnancy does not increase the local recurrence rate after surgical resection of desmoid-type fibromatosis

Justin. M. M. Cates



Adjusted for age, anatomic site, resection margin status and XRT

Table 3 Multivariate analysis of recurrence-free survival of desmoid-type fibromatosis

	Hazard ratio (95 % CI)	P
Pregnancy-associated DTF ^a	1.04 (0.34–3.20)	0.940
Male ^a	1.59 (0.70–3.63)	0.268
Age at DTF diagnosis	0.99 (0.97–1.00)	0.197
Anatomic site ^b	1.80 (0.76–4.29)	0.184
Surgical resection margins ^c	3.71 (1.06–13.0)	0.041

Desmoid-Type Fibromatosis and Pregnancy

A Multi-institutional Analysis of Recurrence and Obstetric Risk

Marco Fiore, MD,* Sara Coppola, MD,† Amanda J. Cannell, BScH,‡ Chiara Colombo, MD,*
 Monica M. Bertagnolli, MD,§ Suzanne George, MD,¶ Axel Le Cesne, MD,|| Rebecca A. Gladdy, MD, PhD,‡
 Paolo G. Casali, MD,** Carol J. Swallow, MD, PhD,‡ Alessandro Gronchi, MD,* Sylvie Bonvalot, MD,†
 and Chandrajit P. Raut, MD, MSc§

TABLE 2. Patients' Characteristics, Treatment, and Outcome According to DF-Pregnancy Relationship

	Group A	Group B	Group C	Group D
No. patients	24	20	29	19
Primary site, n (%)				
Abdominal wall	18 (75)	16 (80)	14 (48)	14 (73)
Extremity	1 (4)	2 (10)	11 (38)	2 (11)
Visceral	4 (17)	2 (20)	1 (3)	2 (11)
Other	1 (4)	0	3 (10)	1 (5)
Primary/recurrent, n/n	24/0	20/0	19/10	17/2
Median tumor size at diagnosis (range), cm	7 (2–35)	6 (3–15)	4 (2–25)	5 (1–19)
Progression during or after pregnancy, n (%)	17 (71)	7 (35)	16 (55)	4 (21)
Treatment after progression, n (%)	13 (54)	7 (35)	8 (28)	3 (16)
Surgery	8 (33)	3 (15)	5 (18)	2 (11)
Medical treatment	5 (21)	4 (20)	2 (7)	1 (5)
ILP	—	—	1 (3)	—
DF progression after definitive treatment, n (%)	3 (13)	2 (10)	8 (28)	3 (16)
Spontaneous regression, n (%)	3 (13)	2 (10)	7 (24)	1 (5)
No treatment after initial watchful waiting, n (%)	7 (30)	7 (35)	9 (30)	—

92 pacientes

Desmoid-Type Fibromatosis and Pregnancy

A Multi-institutional Analysis of Recurrence and Obstetric Risk

Marco Fiore, MD,* Sara Coppola, MD,† Amanda J. Cannell, BSCh,‡ Chiara Colombo, MD,*
 Monica M. Bertagnoli, MD,§ Suzanne George, MD,¶ Axel Le Cesne, MD,|| Rebecca A. Gladdy, MD, PhD,‡
 Paolo G. Casali, MD,** Carol J. Swallow, MD, PhD,‡ Alessandro Gronchi, MD,* Sylvie Bonvalot, MD,†
 and Chandrajit P. Raut, MD, MSc§

TABLE 4. Available Data for Counseling in Women Affected by Sporadic DF

New diagnosis of DF during or shortly after pregnancy	
Risk of relapse after complete resection	13%
Risk of progression with watchful waiting	63%
Spontaneous regression	11%
Risk of failure after any first active treatment (initial or delayed until the time of progression)	10%
Overall managed without resection	52%
Pregnancy after previous diagnosis of DF	
Risk of DF recurrence/progression	42%
DF recurrence/progression safely managed with either active treatment or watchful waiting	94%
Multiple lines of active treatments needed for progression	6%
Spontaneous regression was described after progression as well	7%
Obstetric risk	
Obstetric complications related to DF in both mother and fetus	0%
Intra-abdominal/pelvic DF should be anyway considered at higher risk (few data available)	
Cesarean delivery to be considered in case of macroscopic DF in particular anatomic sites	
Postpartum incisional hernia after previous abdominal wall full-thickness mesh repair is an issue	

Tumeurs Desmoïdes et Grossesse

- Pendant la grossesse 50% de TD progressent

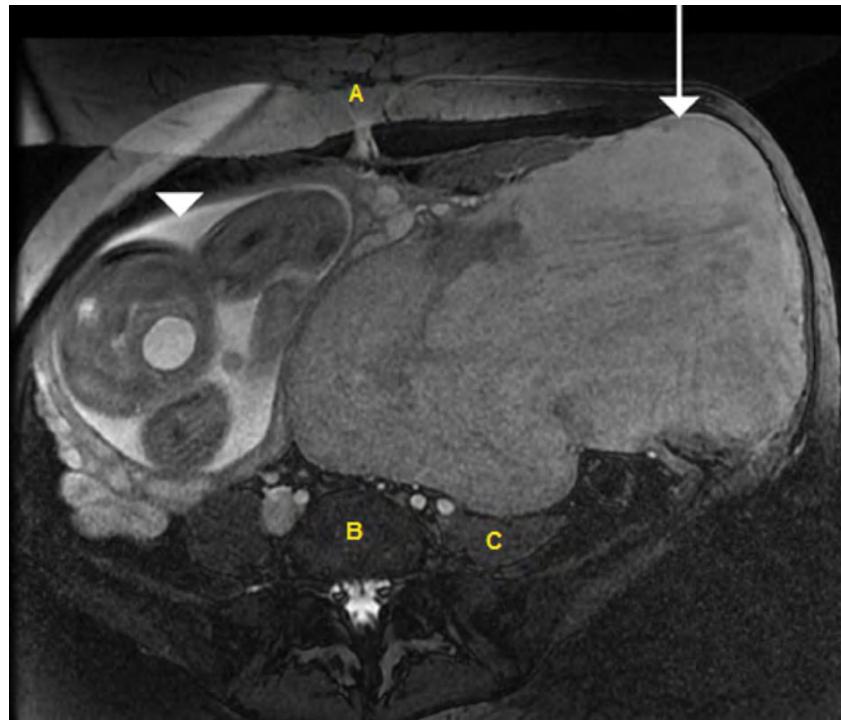


Image: Leon MG et al. A rapidly growing abdominal mass: desmoid tumor in pregnancy. AJR Rep 2015

Tumeurs Desmoïdes et Grossesse

- Surveillance mensuelle par ECHO



- Après l'accouchement 2/3 des TD régressent

Tumeurs Desmoïdes et Grossesse

- TD: pas de contre-indication à la grossesse
- Confirmer le caractère indolent de la tumeur:
attendre 1 an après le diagnostic
- Pas d'indication à une IVG



Merci

