

# A nationwide survey of **Desmoid Tumours** **Management** after labeling of 2 expert networks

## **Nicolas PENEL**

Jean-Michel COINDRE, Axel LE CESNE, Antoine ITALIANO, Agnès NEUVILLE, Sylvie BONVALOT, Philippe TERRIER, Isabelle RAY-COQUARD, Dominique RANCHERE-VINCE, Yves-Marie ROBIN, Nicolas ISAMBERT, Chrisine CHEVREAU, Florence DUFFAUD, François BERTUCCI, Maria RIOS, Maud TOULEMONDE, Cécile LE PECHOUX, Cécile GUILLEMET, Jean-Baptiste COURREGES, Jean-Yves BLAY



# Desmoid tumour – Aggressive fibromatosis

- Rare tumour
- Benign condition – fibroblastic proliferation
- $\approx$  15% related to FAP
- Peak of incidence at 30 – Female: 80%
  
- Difficult pathological diagnosis
- Unpredictable course
- Complex management

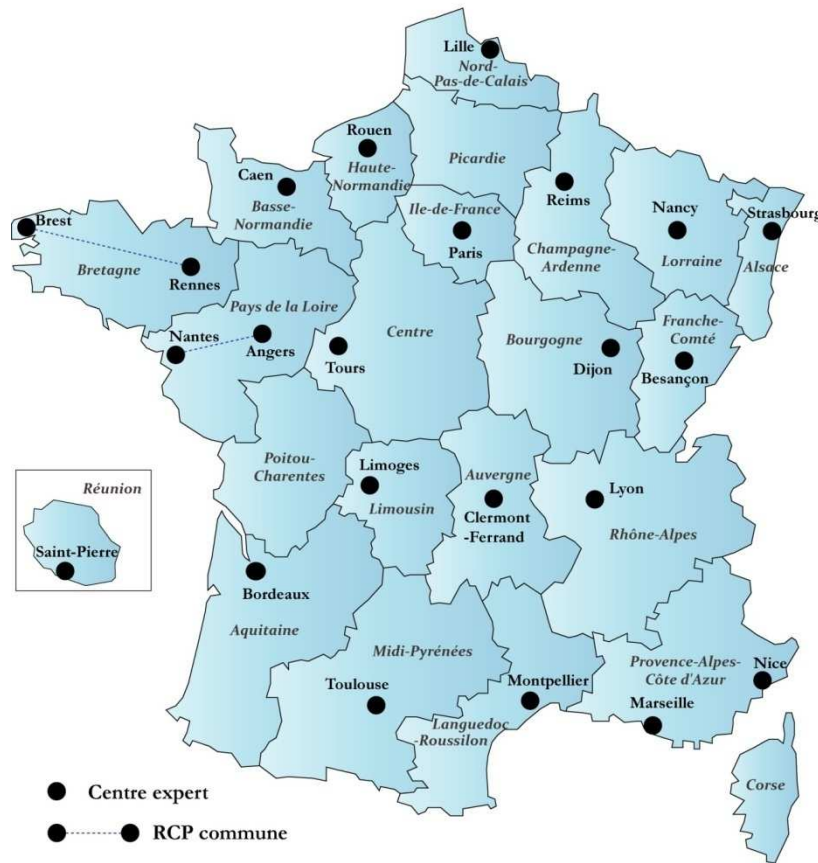
# Labeled Sarcoma Networks

- Initiative of French Cancer Institute Initiative (2009)
- **Without financial or regulatory constraints**
- **RRePs (Pr JM Coindre)**
  - Systematic second pathological opinion by experts
  - Confirmatory molecular biology tests
- **NetSarc (Pr JY Blay)**
  - Clinical management -
  - Dedicaced tumour board
- **Shared data-base - Sarcoma, GIST, Desmoid Tumours (30 000 pts)**



# Labeled Sarcoma Networks

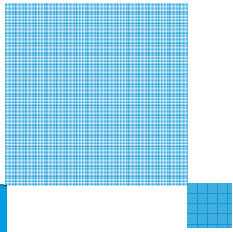
NetSarc  
Réseau clinique des centres experts sarcomes



■ Country Coverage

■ 20 Centres

■ 66 millions of inhabitants



# Labeled Sarcoma Networks

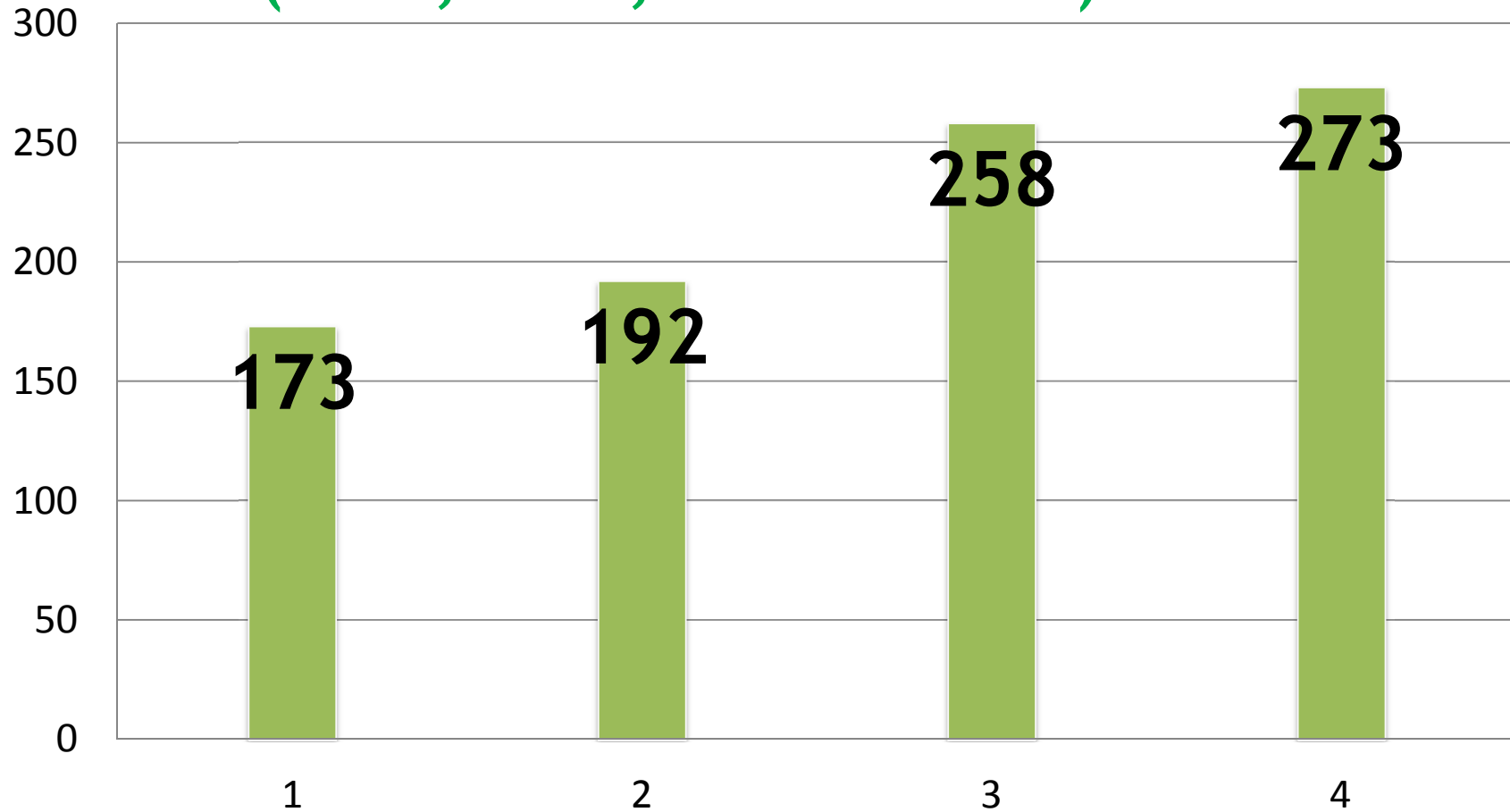
- Analysis of the activity of both networks
- From 2010 to 2013
- Key-indicators of activity



# Activity of pathologist experts

- 861 patients
  - Age from 6 to 90 (median: 44) – *15 cases in children*
  - 573 women (66.6%) and 288 men (33.4%)
- Size from 1 to 55 cm (median: 5.5)
- Most common primary sites
  - 264 abdominal wall 30.6%
  - 188 intra-abdominal 21.8%
  - 116 chest wall 13.4%
  - 33 shoulder 03.8%
  - 33 breast 03.8%
  - 28 thigh 03.2%

# Desmoid tumours confirmed by the pathologist experts (2010, 2011, 2012 et 2013)



Increased of confirmed cases + 57%



## Percentage of cases diagnosed in referral centers

- 2010: 80/173 (46.2%)
- 2011: 65/192 (33.8%)
- 2012: 90/258 (34.8%)
- 2013: 81/273 (29.7%)

**Most of DT have been diagnosed outside Referral centers and then confirmed by expert pathologists**



## Referral centers versus other hospital

	Referral centers <i>n</i> =316	Outside the network <i>n</i> =545	p
Core needle biopsy	195 (61.7%)	120 (22.0%)	>.0001
Surgical biopsy	22 (6.9%)	62 (11.3%)	.76
Surgical specimen	83 (26.3%)	349 (64.0%)	>.0001
Not done	16 (5.0%)	14 (2.5%)	.80

**Surgical specimen: 64% outside referral centers**

**Core needle biopsy: 62% in referral centers**

## Impact of second opinion

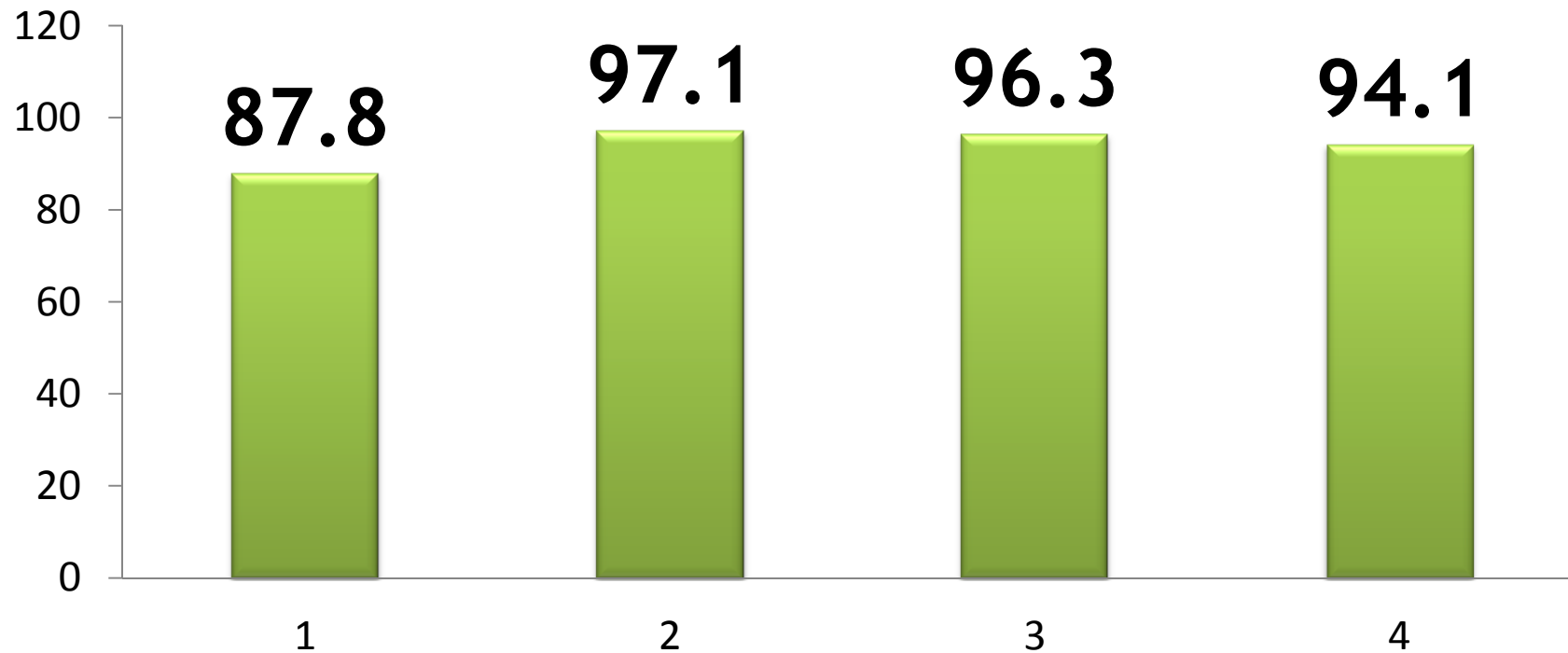
- 561 cases reviewed by experts
- Initial diagnosis
  - Desmoid tumours 405 (72.1%)
  - Other diagnosis 156 (27.9%)
- Most common misdiagnosis
  - Sarcoma 35 (22.4%)
  - GIST 26 (16.6%)
  - Fasciitis 20 (12.8%)
  - Leiomyoma 06 (3.8%)

**Outside referral centers, 1/3 of DT are misdiagnosed**



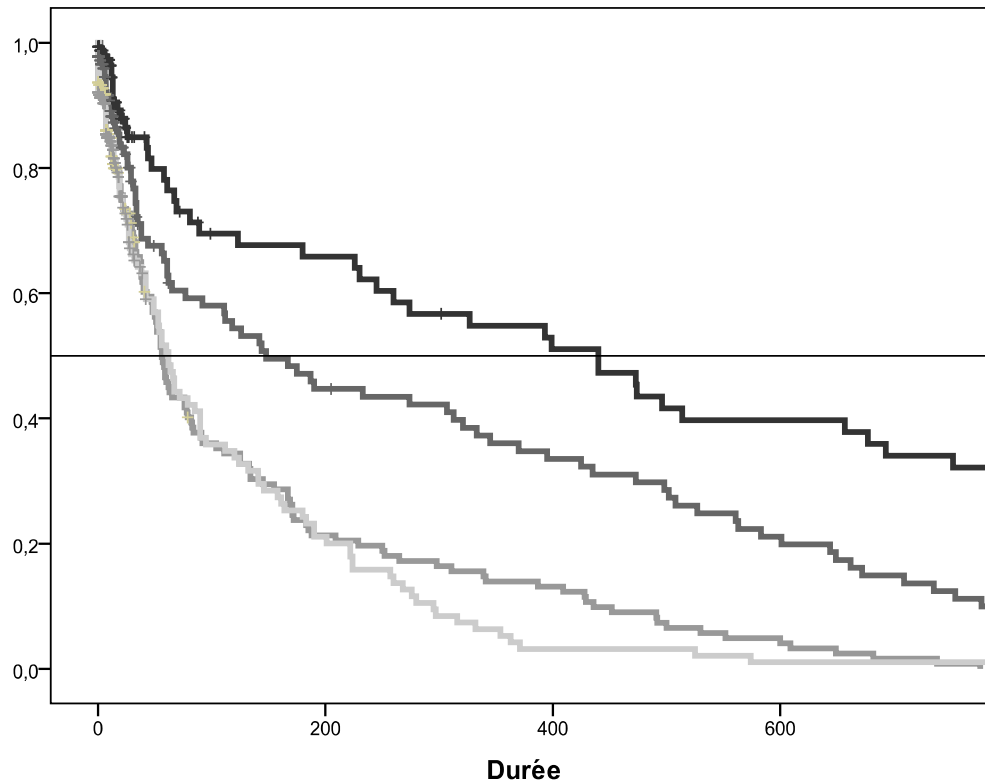
# Percentage of cases with analysis of beta-catenin mutation

(2010, 2011, 2012 et 2013)



<b>Presence of Beta-catenin mutation</b>	<b>583/781 (74.6%)</b>
<b>Absence of Beta-catenin mutation</b>	<b>157/781 (20.1%)</b>
<b>Not analyzable</b>	<b>39/781 (5.3%)</b>

# Median time to diagnosis confirmation (2010, 2011, 2012 et 2013)



▪ 2010: 440 days

▪ 2011: 148 days

▪ 2012: 56 days

▪ 2013: 62 days

$P < 0.0001$



## Activity of clinical centers

- 846 confirmed cases of adult DT
- 414 partly managed in clinical referral centers (48.9%)
- Reasons:
  - Initial management : 207 (50.0%)
  - Second opinion: 160 (38.6%)
  - Relapse or progressive disease: 47 (11.4%)



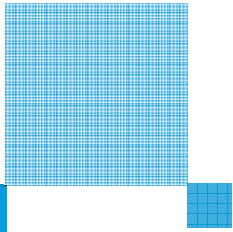
# Factors associated with management in clinical referral centers

	Not seen in referral centers	Managed referral centers	in p
Age (years)	48 +/- 4	<b>44 +/- 4</b>	<b>0.005</b>
Size (mm)	94 +/- 7	70 +/- 7	0.79

	Not seen in referral centers (%)	Managed in referral p centers (%)	p
2010	104 (63.1)	<b>61 (36.9)</b>	<b>0.004</b>
2011	91 (50.0)	91 (50.0)	
2012	106 (44.9)	130 (55.1)	
2013	131 (50.0)	<b>132 (50.0)</b>	
Women	267 (50.0)	<b>298 (50.0)</b>	<b>0.016</b>
Men	165 (58.8)	116 (41.2)	
Bone DT	2 (66.4)	1 (33.3)	<b>0.02</b>
Soft tissue DT	357 (49.2)	<b>369 (50.8)</b>	
Visceral DT	73 (62.4)	44 (37.6)	

	Not seen in referral centers (%)	Managed in referral centers (%)	p
Abdominal wall	128 (49.5)	131 (50.5)	0.82
Other primaries	286 (50.3)	283 (49.7)	
No Beta-catenin M	302 (56.1)	278 (47.9)	0.316
Beta-catenin M	84 (53.5)	73 (46.5)	





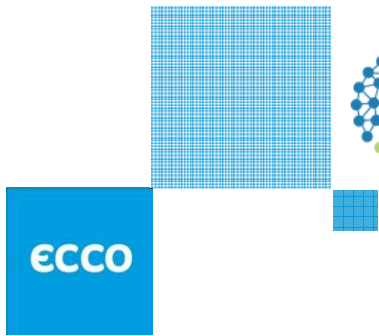
## Conclusion (1/2)

- 1/3 of DT are not correctly diagnosed outside referral centers
- Most of DT diagnosed outside referral centers are operated
- Impact of set-up of pathologist network
  - Increasing in confirmed cases (from 173 to 273/y)
  - Beta-catenin mutation analysis available in more than 95% of cases
  - Decreasing in diagnosis confirmation delay (from 440 to 62 days)



## Conclusion (2/2)

- Increasing in number of pts partly managed in clinical referral centers (36 to 50%)
- Factors associated with management in referral centers:
  - Year of diagnosis
  - Female gender
  - Young age
  - Soft tissue location
- **Impact on outcome needs to be measured**



# Thank you

- Patients and families
- Members of the French Sarcoma Group
- CRA of the French Sarcoma Group, F Ducimetiere
- Patient advocacy Group SOS Desmoïde
- INCa, the French Cancer Institute
  
- No Disclosure

