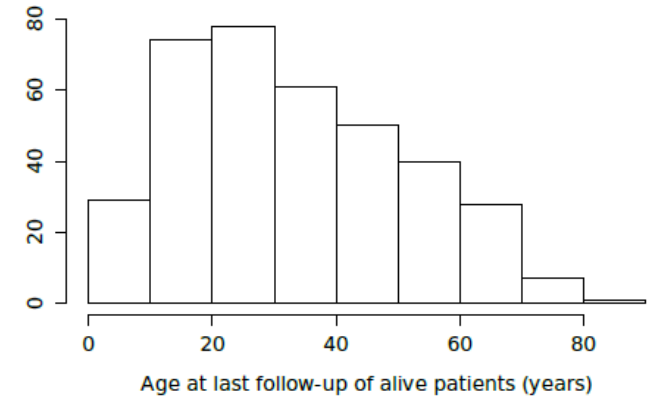


Pregnancies in women with TGFB1 / TGFB2 mutations

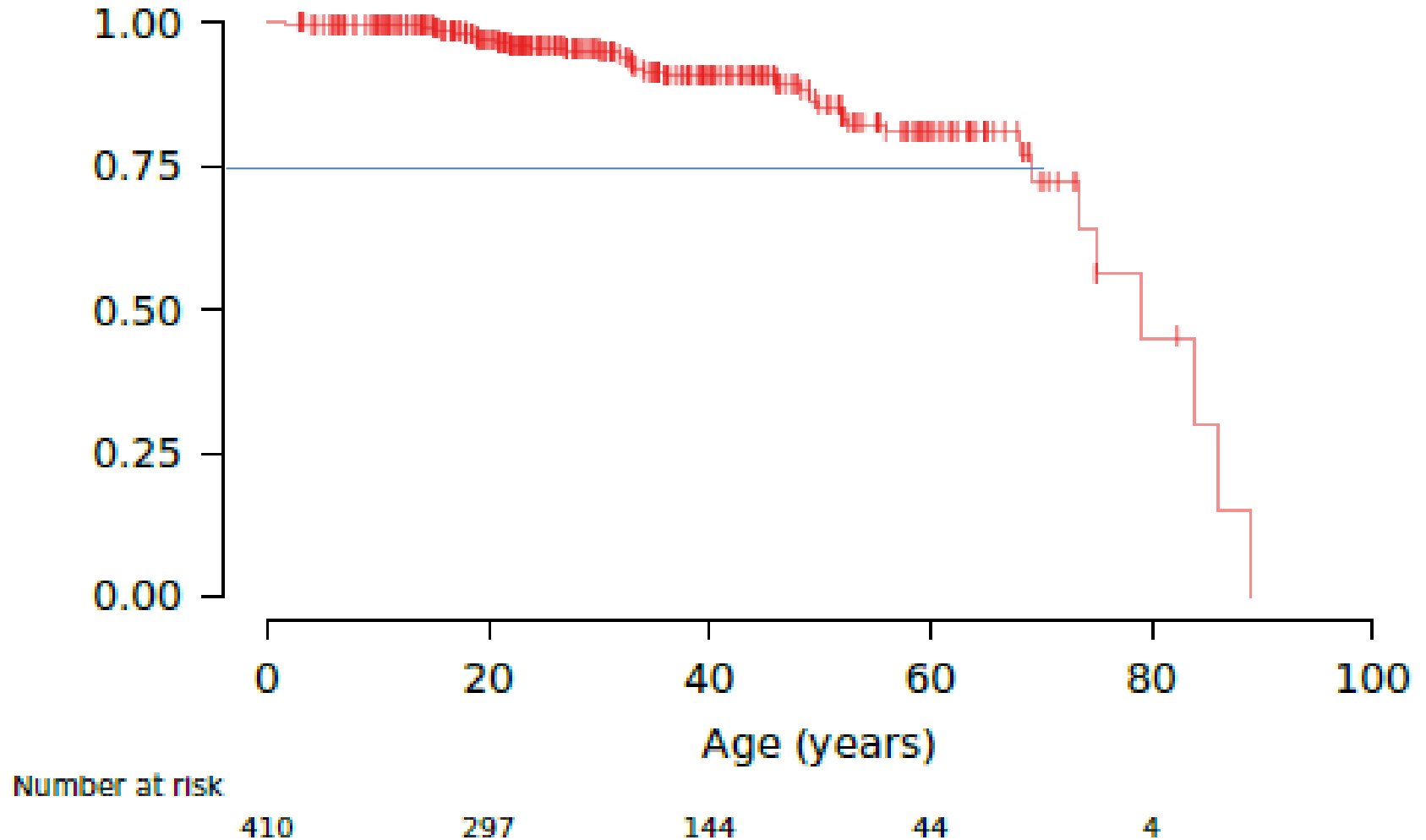
G Jondeau

Population

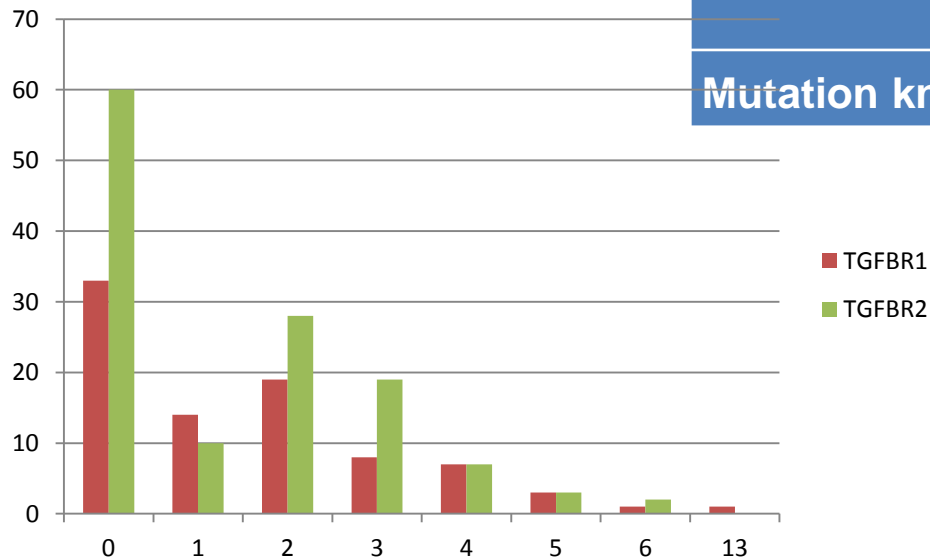
	TGFBR1	TGFBR2	p
N	n=176 (40%)	n=265 (60%)	
Females	99 (56%)	138 (52%)	
Presenting feature			
<i>Aortic root aneurysm</i>	31 (18%)	62 (23%)	
<i>Ao dissection</i>	28 (16%)	40 (15%)	
<i>Dysmorphism</i>	16 (9%)	23 (9%)	
<i>Familial screening</i>	100 (57%)	137 (52%)	



Survival in the whole population



	TGFBR1	TGFBR2	Total
Women with pregnancy	53/99	69/138	122
N° pregnancy	138	178	316
Ao dissection	1 (0.7%)	4 (2.2%)	5 (1.6%)
1st pregnancy	1	1	2
2nd pregnancy		2	2
3rd pregnancy		1	1
Type			
A	1 (surgery)	2 (1 surgery)	
B	0	2 (1-3)	
Mutation known	Y (BB)	N (1BB)	



Conclusion

- 1 pas si grave
- 2 chirurgie précoce marqueur de risque ?
- 3 signes de LDS marqueurs de risque ?

	Dissec	No dissec
Head and neck arterial tortuosity	100% (4/4)	46% (22/48)
Hypertelorism	100% (4/4)	23% (16/69)
Translucent skin	75% (3/4)	56% (58/86)
Broad or bifid uvula	75% (3/4)	22% (17/79)
Arched palate	100% (4/4)	44% (34/78)
% systemic score 7 or greater	¼ (25%)	8/73 (11%)