

# Pre-pregnancy risk assessment and combined multidisciplinary care improves pregnancy outcomes in women with Turner's syndrome

## INTRODUCTION

- There are increasing rates of pregnancy in women with Turner's syndrome (TS), either unassisted (5-5.6%)<sup>1,2</sup>, or with oocyte donation (OD).
- TS is associated with 25-50% cardiovascular malformations<sup>3</sup>, commonly bicuspid aortic valve, dilatation of the aortic root and ascending aorta and aortic coarctation.
- Increased cardiovascular demand during pregnancy raises concern regarding progression of aortopathies, with potential fatal consequences such as aortic dissection.
  - The literature quotes a 2.0-3.3% risk of aortic dissection; 8.0% pre-term birth rate, 3.8% major birth defects and 2.3% perinatal mortality rate in TS pregnancies<sup>3,4</sup>.
- Our local experience in a centre with multidisciplinary management of a large cohort of women with TS was a low rate of maternal and foetal complications.
- The aim of this study was to demonstrate the low complication rate in this cohort of women using a formal follow-up protocol.

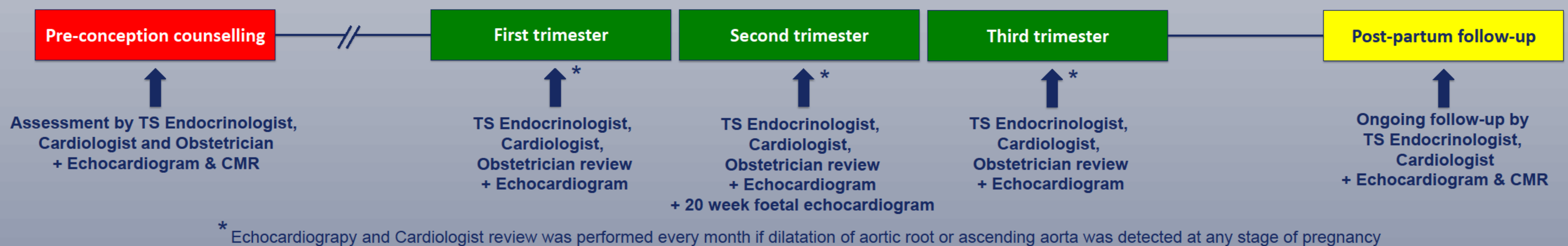
## METHODS

- A database of 101 women with TS from a single specialist unit was retrospectively analysed to identify women with TS and pregnancies.
- Women were grouped as follows:
  - Spontaneous pregnancy with successful gestation and delivery
  - Spontaneous pregnancy with miscarriage
  - Assisted reproduction with OD with successful gestation and delivery
  - Assisted reproduction with OD with miscarriage
- Women had been assessed and treated according to the protocol in Fig 2.
- Blood pressure was strictly controlled prior to, and during pregnancy.
- Echocardiography and cardiac magnetic resonance imaging (CMR) data were analysed to determine aortic root and ascending aortic dimensions prior to, during and after pregnancy.
- Maternal and foetal cardiac complications and mortality data were analysed.



Fig 1. Photo of woman with TS during pregnancy (left), and her daughter (right). Images provided by patient and reproduced with her consent.

Fig 2. Protocol for pre-pregnancy risk assessment and counselling, and monitoring during pregnancy and post-partum by a multidisciplinary specialist team



## RESULTS

	No. of women
Spontaneous pregnancy with successful delivery	12 (n=145XO)
Spontaneous pregnancy with miscarriage only	2
Oocyte donation with successful delivery	3
Oocyte donation with miscarriage only	3
Age at first pregnancy (mean ± SD)	25.5 ± 5.6 years
Number of successful deliveries per woman (median, range)	1 (1-3)
Genetic profile	
45XO	4
Mosaic	16
Pre-existing cardiovascular disease	
Bicuspid aortic valve	7
Aortic root or ascending aortic dilatation <sup>#</sup>	2
Previous coarct repair, VSD, moderate aortic stenosis	1
Hypertension	6
Comorbidities	
Hypothyroidism	2
Crohn's disease with ileostomy	1
Anticardiolipin Ab positive	1
Bicornuate uterus	1
Weeks of gestation (median, range)	39 (37-42)
Method of delivery:	
Caesarian : vaginal delivery	66% : 33%

<sup>#</sup> Indexed for body surface area: Aortic size index (ASI) > 2.0cm/m<sup>2</sup>, associated with an increased risk of dissection<sup>6</sup>.

	No. of women
Maternal pre-eclampsia	0
Foetal complications	
Ventricular septal defect (small)	1
Turner's syndrome in child	1
Aortic sinus or ascending aortic dilatation	3
Indexed dimensions pre- and post-pregnancy (cm/m <sup>2</sup> ):	(1) 1.77 → 1.88; (2) 1.65 → 1.89; (3) 2.73 → 2.89
Age at pregnancy, no. of pregnancies, hypertension (Y/N)	(1) 29 y, 1, N (2) 17y, 1, N (3) 35y (OD), 1, Y
Maternal aortic dissection	0
Maternal mortality	0

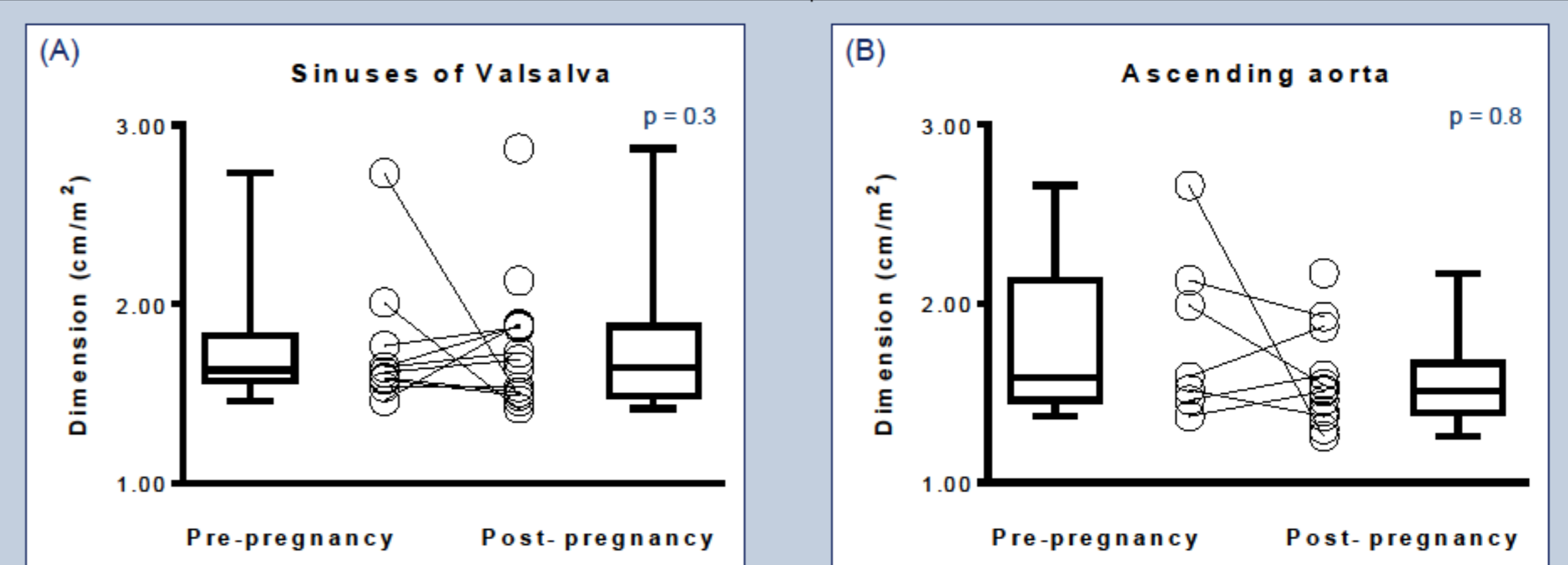


Fig 3. Aortic sinuses (A) and ascending aorta (B) pre- and post-pregnancy (indexed to body surface area, mean, SD, range) p-values obtained using Mann-Whitney unpaired non-parametric test

## CONCLUSIONS

- We noted a higher rate of spontaneous pregnancy with successful delivery (12%), with no adverse maternal cardiovascular complications during pregnancy or post-partum follow-up.
- This protocol of pre-conception counselling and serial follow-up by a specialised multidisciplinary team throughout pregnancy and the post-partum period shows encouraging results, enabling women with TS to undergo safe pregnancy and delivery.
- We suggest that women with TS should be encouraged to discuss pregnancy at an early stage.
- A multidisciplinary approach may improve pregnancy outcomes in these women.