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

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INTRODUCTION

- There are increasing rates of pregnancy in women with Turner’s syndrome (TS), either unassisted (5-5.6%)\(^1\), or with oocyte donation (OD).
- TS is associated with 25-50% cardiovascular malformations\(^2\), 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 aortopathy, 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\(^3,4\).
- 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 only
  - 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.

RESULTS

<table>
<thead>
<tr>
<th>No. of women</th>
<th>Spontaneous pregnancy with successful delivery</th>
<th>Spontaneous pregnancy with miscarriage only</th>
<th>Oocyte donation with successful delivery</th>
<th>Oocyte donation with miscarriage only</th>
<th>Age at first pregnancy (mean ± SD)</th>
<th>Number of successful deliveries per woman (median, range)</th>
<th>Genetic profile</th>
</tr>
</thead>
<tbody>
<tr>
<td>12 (n=1,450)</td>
<td>2</td>
<td>3</td>
<td>3</td>
<td>55.5 ± 5.6 years</td>
<td>1 (1-3)</td>
<td>4500 (4000-5000)</td>
<td>4</td>
</tr>
<tr>
<td>2</td>
<td>16</td>
<td>1</td>
<td>7</td>
<td>1</td>
<td>2 (4-10)</td>
<td>4500 (4000-5000)</td>
<td>16</td>
</tr>
<tr>
<td>2</td>
<td>2</td>
<td>1</td>
<td>1</td>
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</tr>
</tbody>
</table>

Pre-existing cardiovascular disease

- Bicuspid aortic valve: 7
- Aortic root or ascending aortic dilatation: 2
- Previous coarct repair, VSD, moderate aortic stenosis: 1
- Hypertension: 6

Concomitants

- Hypothyroidism: 2
- Cushing’s disease with acromegaly: 1
- Anticardiolipin Ab positive: 1
- Bicuspid aortic valve: 1

Weekly of gestation (median, range) 39 (37-42)

Method of delivery:

- Cesarean: vaginal delivery: 85%, 27%

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.

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

Pre-conception counselling

First trimester

TS Endocrinologist, Cardiologist, Obstetrician review + Echocardiogram + 20 week foetal echocardiogram

Second trimester

TS Endocrinologist, Cardiologist, Obstetrician review + Echocardiogram + 20 week foetal echocardiogram

Third trimester

TS Endocrinologist, Cardiologist, Obstetrician review + Echocardiogram + 20 week foetal echocardiogram

Post-partum follow-up

Ongoing follow-up by TS Endocrinologist, Cardiologist, Obstetrician review + Echocardiogram + CMR

* Echocardiography and Cardiologist review was performed every month if dilatation of aortic root or ascending aorta was detected at any stage of pregnancy.

Fig 3: Aortic stenoses (A) and ascending aortas (B) pre and post pregnancy (indexed to body surface area, mean ± SD, range)

Fig 4: Reference

References:

Poster Presented at ECE 2016 Conference.