## Patients and Methods

## Selection of patients

Cross sectional study included fifteen pediatric patients with TS presenting to the pediatric endocrinology clinic, Cairo University through a period of six months.

The population of the study included age group ranging from 5 to 18 years.

**Inclusion criteria**

* Pediatric Patients with TS (all karyotypes).
* Age group 5-18 years.

**Exclusion criteria**

* Patients were excluded if they have any one of the following criteria: Malignancy, liver disease and contraindications to cardiac magnetic resonance (Including mechanical aortic valve prostheses).

**Interventions**

Study population was studied using cardiac magnetic resonance in order to determine the frequency of cardiovascular anomalies and to assess the utility of this imaging modality as a method for cardiovascular evaluation in TS.

1. Collecting medical data as regards personal data, physical features, percentiles, standard deviation score, blood pressure in the four limbs and detailed cardiological assessment including history of cardiac complaints, general and local cardiac examination to detect possible cardiovascular complications.
2. The parameters that had been assessed by cardiac magnetic resonance include aortic structure, elongation of the transverse aortic arch, aortic diameter at multiple locations, and coarctation of the aorta.
3. 2D Echocardiography had been done for all patients to detect any structural cardiac abnormality, and to assess the ability to detect Aortic abnormalities.
4. Associations between data obtained from the cardiac magnetic resonance and the following factors were assessed: age, karyotype, phenotype, body surface area, blood pressure and 2D echocardiography.

**Methodology**

After obtaining an informed consent from patient`s guardians, Patients enrolled in the study were subjected to:

* **Full history taking** including personal data, consanguinity, cardiac complaints.
* **Thorough clinical assessment** including measurement of height, weight, surface area and body mass index, blood pressure measurement, full systematic examination with special emphasis on dysmorphic features, blood pressure in the four limbs and detailed cardiological examination and possible cardiovascular complications.
* **Investigations:**

1. **Laboratory**:

Karyotyping for all included cases was documented.

1. **Imaging:**

* 2D Echocardiography for: detection of structural cardiac abnormalities including valvular lesions, septal and wall abnormalities, in addition to vascular abnormalities as Aortic coarctation or dilatation.

The examination was performed using General Electric, Vivid 3 Pro, Sync Master 591S device with 3.5-7 MHz probe. All cases were examined by the same operator.

* Cardiac magnetic resonance imaging for:

Detection of delicate abnormalities in the heart and Aorta. Data collected from CMR includes the diameter of:

1. Aortic annulus.
2. Bisinus diameter.
3. Sinutubular junction (STJ).
4. Ascending Aorta.
5. Aortic arch.
6. Descending Aorta.

The MRI device used in the study is Gyroscan 1.5 Tesla.

**Interpretation of data**

We calculated the aortic ratio which is the ascending aortic diameter in relation to descending aortic diameter (AD/DD). Ascending aorta was considered dilated when this ratio was more than 1.5

The ascending aortic diameters were correlated in relation to body surface area to calculate the aortic size index (ASI) which is the ascending aortic diameters normalized to body surface area.

According to ASI, patients were classified into:

* Patients had ASI values greater than 99th percentile (2.5 cm/m2) were listed as severe dilatation for intervention because they have high risk of aortic dissection.
* Patients had ASI values greater than the 95th percentile (2.0 cm/m2) were listed as moderate dilatation for follow up every 6 months**.**
* Patients had ASI values approaching the 95th percentile (2.0 cm/m2) were listed as mild dilatation for follow up CMR after 3-5 years **(Carolyn and Bondy, 2008).**

**Table (4- 11). Significance of Aortic size index (ASI)**

|  |  |  |
| --- | --- | --- |
| **Normative** | **ASI** | **Clinical concern** |
| 95th centile female controls | >2.0 cm/m2 | Close monitoring |
| 99th centile female controls | ≥2.5 cm/m2 | Evaluate for intervention |

ASI = ascending aortic diameter at the pulmonary artery origin / body surface. **(Carolyn and Bondy, 2008).**

**Statistical analysis**

Data were tabulated and subjected to computer-assisted analysis using Microsoft Excel version 2010 and the Statistical Package for Social Science (SPSS) version 21.

The following methods were employed:

* Frequency distributions and percentage distributions.
* Mean, standard deviation, median, range and interquartile ranges of numerical data.
* Comparison of quantitative variables using the Mann-Whitney test.
* Comparison of qualitative valuables using chi-square with Fisher’s exact test.
* Spearman correlation coefficient was conducted to signify the association between different quantitative variables.
* Kappa measurement of agreement was calculated to test the concordance between ASI and ARI.
* P values less 0.05 was considered significant.
* Graphs illustrated some information.
* Charts were prepared using Microsoft Excel 2010.