Features and Concepts: Saudi Congenital Heart Defects Registry

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Multi-Institutional Registrar
Congenital Heart Defects Registry
Congenital Heart Defects Registry (CHDR) observes an ongoing process of data collection, entry, analyses and reporting for a defined population diagnosed with Congenital Heart Defect(s).

The registry is an approved research collaborative project since 1998 between King Faisal Heart Institute and Research Center, Riyadh (KFSH&RC).

The registry expanded its collaborative efforts on a multi-institutional level in the year 2003.
Objectives

- To provide leadership in establishing and maintaining comprehensive CHD registration with other health organizations.
- To support scientific and clinical research.
- To provide hospital administrators with sufficient data for proper health planning.
- To serve as a surveillance tool for monitoring long-range trends of congenital heart diseases.
All patients diagnosed to have CHDs are eligible for registration, regardless of their age, gender or nationality.
The European Pediatric Cardiology Coding list (EPCC) is the system in use to code the abstracted disease and interventional descriptions.
The registry is a web based software, with high levels of security:

- Software cookies set-up
- Unique ID and password for each user
- Various users levels
- Tracking system identifying database access

Being an internet application all activities are real-time:

- Data entry, coding, auditing and data download
- Patient transfer
- Report generation
- Chart generation
- Real-time data search
- Blog (communication form for interested community)
CHDR is a centralized real time database. All data is hosted on a secured web-server.

Observing and ensuring the complete data segregation:
- Each collaborating institute has its unique institutional code
- CHDR staff has an access to the centralized data base according to the assigned privileges on their respective institutes
In line with and adhering to the CHDR multi-institutional bylaws, many measures are in place that preserves the confidentiality of the collaborating hospitals and patients by observing:

- Masking of the hospitals' names and codes
- Masking of the Medical Record Numbers
- Masking of all identifiable data of the patients (name, ID #)
Physicians and researchers with browse privileges obtain an up-to-date information through:

- **Search page:** a search engine helping researchers to tailor their queries by selecting available set of parameters for quick data analysis

- **Chart generation:** feature to obtain real-time data as distribution tables and charts
Data reported as of February 5th, 2011
20,223 • Total number of registered patients since the inception of the registry.

13,834 • Active follow up patients as per their latest recorded follow up visit in the database.

4,729 • 16 years and above stated alive according to their latest database recorded follow up.

3,662 • Alive with active follow up as per latest database recorded.

300 • Discharged from the cardiology services follow up of their reporting hospitals.
Distribution of CHD Patients based on Parental Consanguinity and Family History of CHD

First-Cousin Matings and Congenital Heart Disease in Saudi Arabia
Community Genetics; 1999; 2, 2-3; ProQuest Medical Library pg.69 Susan Becker; Zohair Al Halees
Patients within the same families with positive history for CHD are clustered and are assigned a unique family number.

Around 600 families with a congenital heart defects are assigned a unique family number.
Distribution Of CHD Patients - Age at Diagnoses

- Prenatal: 2%
- 0-365 days: 71.60%
- 1 year and above: 26.40%
Life Status of Prenatally Diagnosed CHD Patients

- Alive on active follow up: 69.1, %
- Deceased: 7.3, %
- Discharged: 5.4, %
- Lost to Follow/Not documented: 18.2, %
Gender Distribution - CHD Patients

10,394
51%

9,829
49%

Male
Female
Congenital Heart Defects – Gender Distribution

- Long QT syndrome
- Hypoplastic Left Heart Syndrome
- Congenital Complete Heart Block
- Congenitally Corrected Transposition of Great Arteries
- Pulmonary Valve Atresia
- Aortic Valve Stenosis
- Coarctation of Aorta
- Transposition of Great Arteries
- Patent Ductus Arteriosus
- Pulmonary Valve Stenosis
- Tetralogy of Fallot

Pink bar = Female  
Blue bar = Male
9,266 patients underwent 14,366 episodes of heart surgery
Distribution Of Heart Surgery Episodes According to Patients’ Age At Intervention

Data reported as of February 5th, 2011
Total of 659 patients underwent Fontan procedure.

Results are displayed in serial number no hospital identification.
Total of 659 patients underwent Fontan procedure distributed as follows:

**Total of 648 CHD Patients underwent Fontan procedure at age less than 16 years**
- 596 Alive
- 36 Deceased
- 7 Discharged
- 21 Lost to Follow

**Total of 14 CHD Patients underwent Fontan procedure at age above 16 years**
- 10 Alive
- 1 Deceased
- Non
- 3 Lost to Follow
Hypoplastic Left Heart Syndrome

- 54 patients above age of one year underwent different heart surgeries
  - 42 patients are alive on active follow up.
  - 12 patients deceased

- 134 patients underwent different heart surgeries under age of one year
  - 68 patients are on active follow up
  - 47 patients deceased
Extra-Cardiac Health Problems

- Down's Syndrome: 1653
- William's Syndrome: 73
- Noonan's Syndrome: 41
- Marfan's Syndrome: 37
- Digeorge's Syndrome: 27
A pilot study is conducted at KFSHRC Riyadh on a selected sample of CHDs patients.

Cases are randomly selected based on the Risk Assessment Congenital Heart Surgery (RACHS).

Average cost is estimated for data pertaining to:

- Different cardiac investigations (2-D Echo, diagnostic cardiac catheter, ST scan, MRI and others).
- Heart surgery procedures.
- Interventional cardiac catheter.
- Total number of follow up visits.
http://rc.kfshrc.edu.sa/chd_program/