Proceedings of the third meeting of the Birth Defects Registry held on 3rd October 2001

The third meeting of the Birth Defects Registry was held on the 3rd of October 2001 at Mediscan Systems, Chennai. Dr.S.Suresh, Director, Fetal Care Research Foundation (FCRF), gave the welcome address. Dr.Sujatha Jagadeesh (FCRF) extended a hearty welcome to the two new hospitals; Kanchi Kamakoti Child's Trust Hospital (KKCTH) & Devaraj Manikchand Maternity Hospital, who were enrolled as members of the registry.

This was followed by a case presentation by Dr.Kalpana Gowrishankar (KKCTH). She reported two children with possible diagnosis "Winchester syndrome", a rare nonlysosomal connective tissue disorder with autosomal recessive inheritance. It is characterised by short stature, polyarthralgia, progressive joint deformities, flexion contractures of small and large joints, coarse facial features, thickened and hyperpigmented skin, hypertrichosis and peripheral corneal opacities. Excessive abnormal oligosaccharides are found in the urine. These children have normal intelligence.

Radiological features include extensive progressive osteoporosis, thinning of long bones, resorption of distal phalanges, subluxation of cervical vertebrae leading to compression fractures etc.

Dr. Kalpana said ten cases have been reported so far. Possible supportive measures that could be offered to such cases by coordinating with the other affected persons were discussed.

Dr.G.Thangavel (Epidemiologist, FCRF) discussed various issues like, a few modifications made in BDR forms to make them more user friendly and coding of congenital malformations based on ICD-10 published by WHO. He presented preliminary report on the birth defects statistics from the 12 participating hospitals of the registry, over a period of seven months from January 2001-July 2001. FCRF statistics was analysed separately as it is basically a referral prenatal diagnostic center. Details regarding the statistics presented at the meeting are given overleaf. The total prevalence estimated as 18.8/1000. Prevalence of musculoskeletal anomalies constitutes a major proportion of all the anomalies followed by CNS anomalies in the birth defects analysed. Analysis of FCRF statistics revealed the fact that autopsies help us to pick up more intricate cardiac malformations.

The meeting ended with a satisfactory note from Dr.S.Suresh on the progress made by the registry. This meeting had been an eye opener to the prevalence of birth defects within a limited sample population. He hoped that it would spread it's wings shortly covering the whole city and subsequently the other neighboring states. He felt that by extending it to a larger population we could present more authentic data on birth defects and bring about preventive/supportive programs. Efforts are on to provide online registration of birth defects.

FCRF also has made arrangements to enroll our registry in the prestigious International Clearing House For Birth Defects Monitoring, Rome, Italy. Dr.Sujatha Jagadeesh emphasized the need for the paediatric personnel of the member hospitals to contribute information regarding birth defects as our registry includes cases up to one year of age. This contribution will bring to light the prevalence of non-lethal/correctable birth defects. The next meeting is scheduled to hold in the last week of December 2001.

Chennai Birth Defects Registry
A preliminary report (January - July 2001)
Dr.G.Thangavel (FCRF)

Introduction:

A Birth defect registry is a systematic mechanism for the collection, storage and reporting of congenital anomalies on a defined geographical area. In 1993, The Fetal Care Research Foundation, a charitable trust was formed to do further research in fetal malformations, prenatal diagnosis and fetal therapy. As an offshoot of FCRF, The Birth Defects Registry (BDR) was started in the year 1996. In 2001 January it was expanded by inducting 13 more hospitals in its fold.

Methodology:

Chennai Birth Defects Registry is a hospital-based registry, where data are collected on the basis of place of delivery rather than residence of the mother (population based). The Data collection is done in a passive manner i.e. instead of the registry staff going to every participating centre and retrieving data from the medical records, the obstetrician or paediatrician of the participating centre at the time of diagnosing a birth defect, write the details of the demographic,

maternal, obstetric and the congenital malformation(s) of the fetus on a form, which is provided by the registry. This form is then sent to the registry where it is reviewed by the paediatricians / dysmorphologists. All modes of deliveries that took place at the participating centres are enumerated in the registry. They include all the live & stillbirths (LB & SB), Intrauterine deaths (IUD), spontaneous or missed abortions, medical termination of pregnancy (MTP) in view of an anomaly or if the couple opted for the same.

Definitions:

- A congenital malformation is defined when a structural or chromosomal anomaly is found in a live born or IUD/SB or aborted fetus, seen at the time of delivery or within the infants 1st birthday.

- Abortion is defined as any intrauterine fetal loss before 24 weeks of gestation. It is further classified as (a.) Spontaneous/ Missed, (b) Medical termination of pregnancy in view of anomaly (c) Medical termination of pregnancy opted by parents.

- Intra uterine deaths and stillbirths are grouped under one category as any intra uterine fetal demise after 24 weeks of gestation.

Diagnosis of a congenital malformation can be modified / updated if a postnatal finding (Karyotype or Autopsy) showed any additional features / altered diagnosis.

Coding of congenital malformations:

Chennai BDR follows ICD –10, published by WHO (World Health Organisation, Geneva), for coding of congenital malformations. Codes are alphanumeric & have four characters with the prefix 'Q', which represents congenital.

Storage & analysis of Data:

Once the forms are reviewed, each fetus is assigned one unique ID as an identification variable and the data are stored in a computer. All information reported to the registry is held in strict confidence. In the tabulations, a baby/fetus with several anomalies is counted once within each class of anomaly. Therefore, the number in different classes cannot be added to reach a total number of babies/fetuses. Isolated anomalies are not distinguished in the results from anomalies that form part of syndromes or multiple malformations. When a baby/fetus has two or more conditions corresponding to the same code, it is counted once only. Prevalence of each participating center will not be analysed individually. Most registries present tables with several prevalence rates: total prevalence rate (LB, IUD/SB and abortions), prevalence rate in livebirths, prevalence rate in livebirths and IUD/SB and contribution of MTPs (in view of an anomaly) to total prevalence rate. This is performed in order to improve comparability between registries and with other sources either restricted to livebirths or live and stillbirths but excluding MTPs.

However, this first report presents only the total prevalence rate. The calculation of total prevalence is done in the following manner.

$$\text{Total prevalence} = \frac{LB_{cm} + IUD/SB_{cm} + A_{cm}}{\text{Total number of deliveries}}$$

LB_{cm} = *Congenitally malformed live births*

IUD/SB_{cm} = *Congenitally malformed IUD/ Still-births*

A_{cm} = *Congenitally malformed abortuses (Incl. Spontaneous/missed, & MTP - by option & in view of anomaly)*

Total number of deliveries = Total number of (LB + IUD/ SB + Abortions)

Results:

This first report of Chennai Birth Defects Registry (BDR) presents the prevalence of congenital malformations occurring among the 7,544 deliveries, which took place in the 12 participating centres of the registry from January to July 2001. Two centres were excluded from the analysis for the following reasons,

- FCRF – which does not conduct deliveries and

- St. Isabel's hospital – which is yet to report the delivery statistics to the registry

However, their congenital malformation statistics were presented without the calculation of prevalence. Table. A shows the split up of deliveries.

Table A

Type of deliveries	N	%
Live births	6879	91.2
IUD / Still births	119	1.6
Spontaneous / Missed abortions	329	4.4
MTP (Option / Anomaly)	217	2.8
Total deliveries	**7544**	**100**

There were 142 malformed babies among the 7544 deliveries, i.e. the total prevalence is 18.8 per 1000 deliveries. Skeletal anomalies have the highest frequency (40) with the prevalence of 5.30 per 1000 deliveries, followed by CNS with the prevalence of 5.04 (38). Talipes equinovarus (TEV) is the commonly found malformation, 2.65/1000 (20), followed by spina bifida without hydrocephalus 2.12/1000 (16). When the contribution of each system was analysed to the total anomalies, it was found that skeletal anomalies contribute 20%, (Figure-1a) in other words every fifth malformed baby has a skeletal

anomaly, followed by CNS malformations (18%). Fig-1b shows the system specific malformation of the FCRF & St.Isabels Hospital.

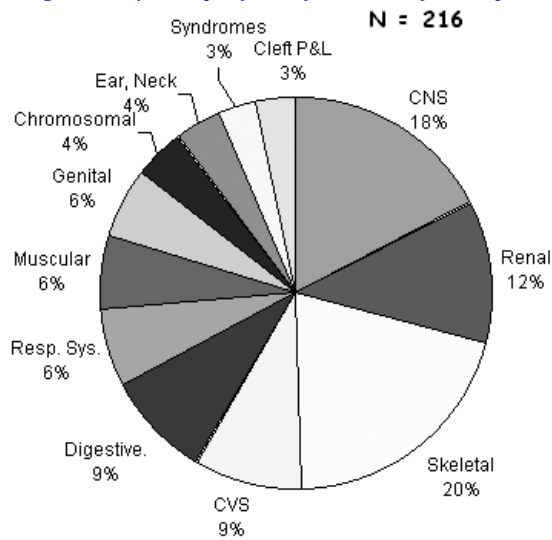*Figure.1a: System specific malformations of 12 hospitals*

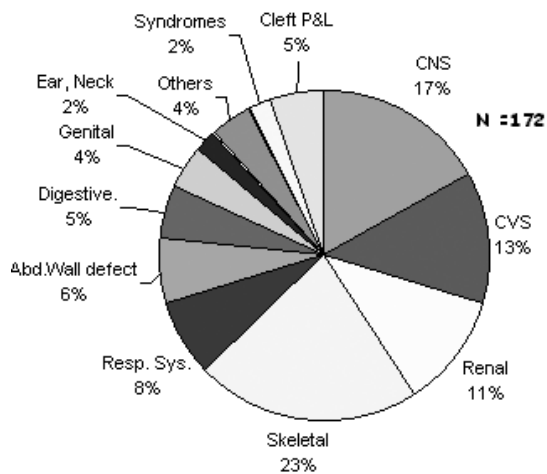*Figure.1b: System specific malformations of FCRF & St. Isabel*

Fig 2 (a & b) show the split up of Musculoskeletal malformations. Talipes equinovarus is the most frequent malformation found. In CNS malformations spina bifida with out hydrocephalus is the most commonly found malformation (Fig.3a & b). When CVS malformation were analysed septal defects constitute about 27% & 41% (Fig.4a & b).

Figure.2a: Musculoskeletal malformations of 12 hospitals
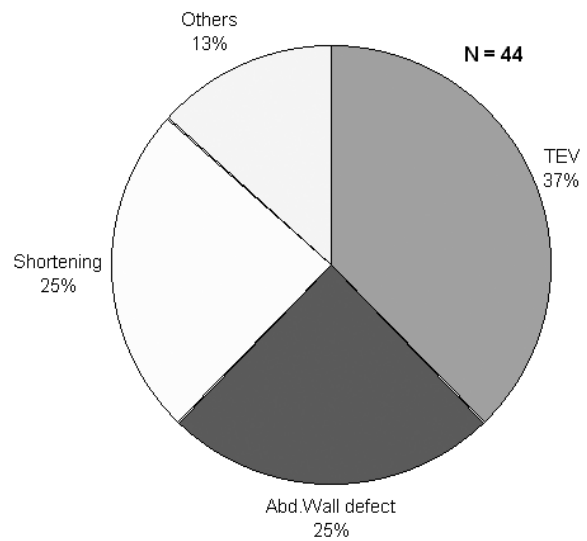
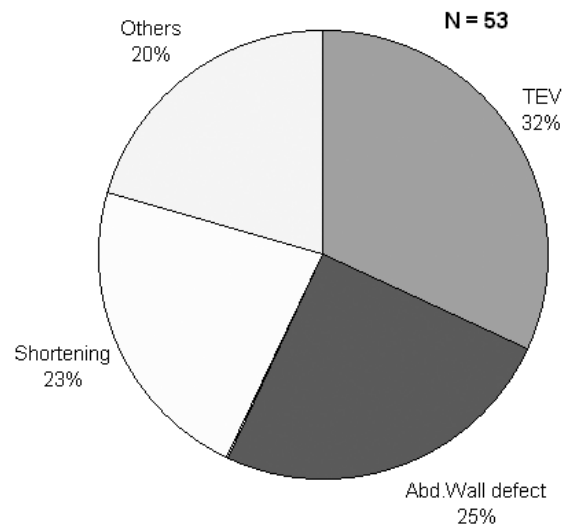*Figure.2b: Musculoskeletal malformations of FCRF & St. Isabel*
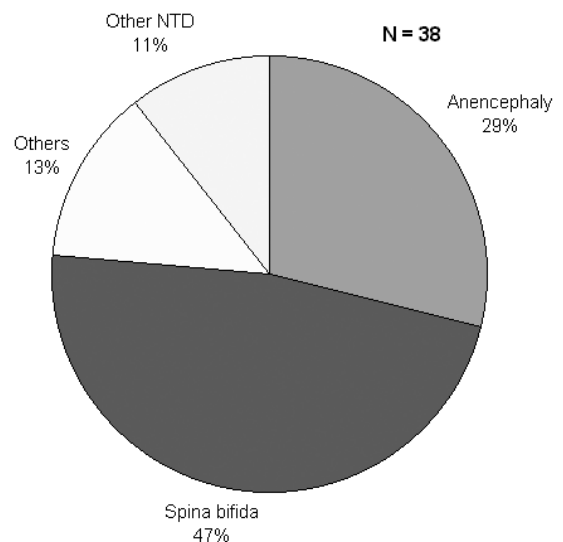

Figure.3a: CNS malformations of 12 hospitals

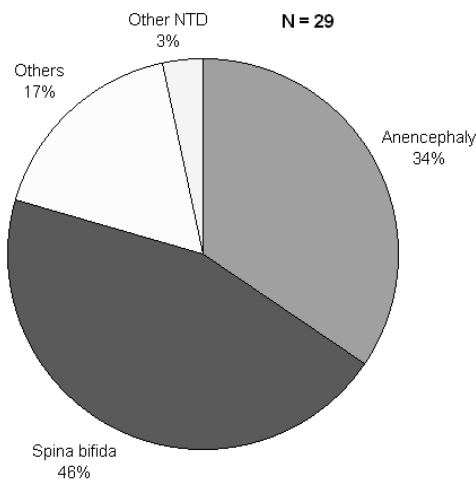*Figure.3b: CNS malformations of FCRF & St. Isabel*

Other NTD 3%
Others 17%
N = 29
Anencephaly 34%
Spina bifida 46%

Figure.4a: CVS malformations of 12 hospitals

HLHS 7%
N = 41
Septal defects 27%
Others 66%
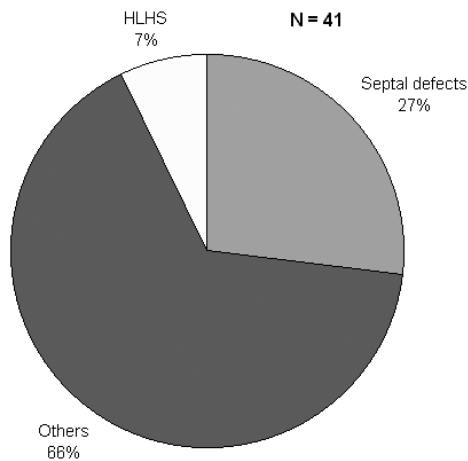
Figure.4b: CVS malformations of FCRF & St. Isabel

HLHS 15%
N = 27
Septal defects 41%
Others 44%
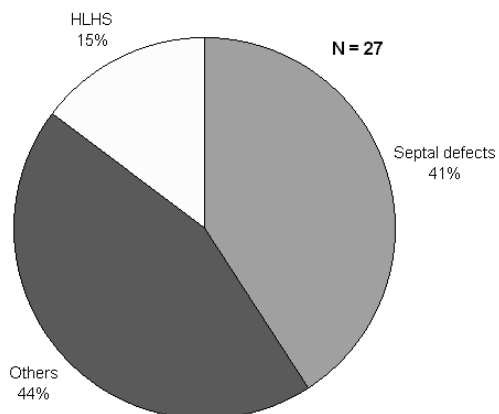
Following is the list of Doctors who represented their Hospitals at the third BDR meeting on 3rd October 2001

Name of the Hospital	Participants	Code
Mediscan Prenatal Diagnosis & Fetal Therapy Centre	Dr. S. Suresh, Dr. Indrani Suresh, Dr. Sujatha Jagdeesh, Dr. Lathaa Bhat, Dr. Gazala Jabeen, Dr.G. Thangavel, Mrs. Ranjani Pathasarathy, Mrs. Chandini Rajendran, Ms. Rehana	001
Kanchi Kamakoti Child Trust Hospital	Dr. Kalpana Gowrisankar	003
Sundaram Medical Foundation	Dr.Bhuvana.S	005
Vijaya Hospitals	Dr. Mona, Dr. Vani Pujari	006
Sri Ramachandra Medical College Hospital	Dr. S. Balagopal, Dr. Usha Viswanath	008
Public Health Centre, West Mambalam	Dr. Prabha Ganapathy,	011
C.S.I. Kalyani Hospital	Dr. Shanthi Durairaj	013
Devaraj Manikchand Maternity Hospital	Dr. Revathi Ravikumar	016

Other Member Hospitals

Name of the Hospital	Code
E.V. Kalyani Medical Centre	002
St. Isabel's Hospital	004
Apollo Hospitals	007
Durgabai Desmukh General Hospital	009
Corporation Hospital, Saidapet	010
CSI Rainy Multi Specialty Hospital	012
Nagamani Hospital	014
G.G. Hospital	015