B D R News

The official newsletter of the Birth Defects Registry of India, (A unit of Fetal Care Research Foundation)

Volume 11 Issues 3 & 4 combined: Sept & Dec 2011

Proceedings of the Third & Fourth meeting of BDRI for the year 2011 was held on 14.09.2011

Dr. Sujatha Jagadeesh gave the introductory speech in which she elaborated on the achievements of BDRI for the year 2011. Dr. Sujatha said that 65 new hospitals had been enrolled, a new State of Sikkim had joined after which BDRI had covered all the States of India and therefore could proudly call itself a National Registry. 19 new Government hospitals had been included and online registration and reporting had become user friendly due to which many hospitals were reporting online. Dr. Sujatha requested all BDRI members to co-operate in improving the quality of the reported data by supporting them with images or pictures which would go a long way in improving the quality and authencity of the registry. She said that BDRI was looking forward to more Doctors and specialists to enroll and to contribute for the national cause.

Lastly she said that the hard work of BDRI had been recognized by its inclusion to the International Clearing House for Birth Defects Surveilance. After enumerating the achievements of BDRI Dr.Sujatha went on to introduce the Chief guest Dr.Sivakumar an eminent cardiologist of Miot Hospitals. Dr.Sivakumar,after his UG went on to complete *DCH,MD,DNB(ped) and DM/DNB(cardio).* With several achievements and awards to his credit his topic for the talk was " Recent Advances In Interventions in Congenital Heart Diseases."

Recent advances in catheter interventions in congenital heart diseases by - *Dr. Sivakumar MD DCH DNB (paed) DM DNB (card) - MIOT hospital*

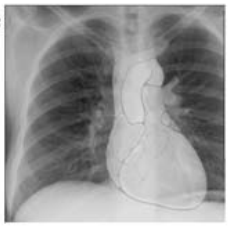
Acyanotic heart diseases	Cyanotic heart diseases
Defects (ASD, VSD) Valvar stenosis Coarctation PA stenosis	Balloon septostomy Pulmonary AV fistula Ductal stenting

A)ACYANOTIC CONGENITAL HEART DISEASES

This includes

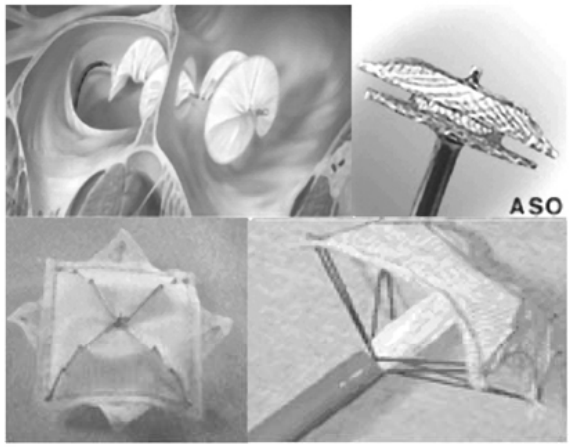
1) **Atrial Septal Defect** {ASD}: In majority of cases about 80% of ASD detected are **secundum ASD.** In a large proportion of secundum ASD **intervention** is possible.

2D Echocardiography - is the investigative procedure used to view and to identify the size of the **defect.** Usually the defects are viewed through



a) parasternal window,
b) apicalwindow,
c) subxiphoid window

Once the defect is identified, the size and the margin of the defect is noted. Then the device is selected. **The device** consists of two discs on either side with a central waist. Once the size of the defect is noted for eg. If the size is 18mm the device selected should have 18mm waist and 6 - 7mm larger retention skirts in either sides, so that when introduced into the defect and pulled up, it grips to the sides and complete closure is acheived. The technique is performed in such a way that the device fits perfectly into the defect. Commonly used device is **amplatzer septal occlude device.**



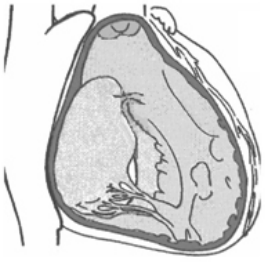
In Patients with multiple defects, 3D echocardiography gives additional spatial orientation of the defects and these multiple defects can be closed by interventions. Around the world,about 2lakh devices have been deployed. The safety of the procedure is well assured. Initially (10yrs back) there were conventional indications for interventions - Such as *Secundum ASD should have RA RV Volume overload and the size of the defect can be only. upto 34mm and with 5mm margin on all sides.* Nowadays, however with experience, defect upto 44mm and even with deficient margins and multiple defects - can be closed safely (multiple devices are used in closure of multiple defects and are done simultaneously). Presently Secundum ASD is being corrected mostly by intervention only, surgery is rarely opted for.

2) Ventricular septal defect (VSD) - In VSD, the common type is the (a) **membranous VSD (about 80%).** VSD is also viewed by 3D Echocardiography. A catheter is passed through Aorta into the Left ventricle and the device is deployed. The double umbrella device used in VSD closure is made up of Nitinol (nickel & Titanium) which has a good thermal memory. The texture of the material is such that, when it is crushed and passed into the defect, it adjusts accordingly and fits in. Here also **Amplatzer membranous VSD occlude device** which is asymmetric, is commonly used.

The other types of VSD are (b)Isolated Muscular VSD {occurs in about 10%} and (c)Inlet and Outlet type VSD - **Inlet type is AVcanal VSD and Outlet type is Sub pulmonary VSD** both contribute 8-10%. The inlet and outlet types are not amenable because they are close to pulmonary or Avvalves.

Majority of muscular VSDs can be closed interventionally. The main criteria is that, the Baby's wt should be 10kg and above for intervention.When the baby is weighing less, with associated heart failure, recurrent lung infections, growth failure, ***Surgery is advisable*** rather than waiting for the child to grow to a weight where intervention can be performed.

Ventricular septal defect

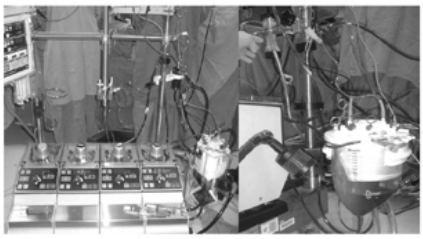


Proceeding from the ventricles to the ascending aorta, the connection between ascending aorta and pulmonary artery is the Distal Aorto Pulmonary window and the connection between of arch of aorta and the pulmonary artery is the **Patent Ductus Arteriosus (**PDA):

PDA closure was the earliest intervention to come. PDA coil closure was started in 1980's. Initially the coils used were made of stainless steel or platinum.They contains dacron fibres and these fibres facilitate thrombosis around the coils. Once the coil is deployed-the dacron fibres enhance the thrombogenicity around the same and forms a complete closure of the defect. Initially, single coil was only used. Later, 3 - 4 coils tied together with Bioptome-were used.In recent times **Amplatzer device** is the commonly used device. Even upto 25mm of defect are closed by interventions. PDA closure is almost always corrected by intervention instead of surgery.

Ductus in prematurity is considered to be challenging for both, the cardiologist and the cardiac surgeon. When the child is not responding to NSAIDS (which is used to suppress prostaglandin mediated ductal patency), Intervention **or** Surgery is opted for. **In surgery** complications arise in the post operative period like lung collapse, pneumothorax, or sepsis. **Intervention** method is used in selected PDA with acceptable morphology {Acceptable morphology means the size of ductus should be less than 3-4mm} Though **Interventions** also have complications, the recovery is very fast.

Dr.Sivakumar explained about the reasons for preferring catheter intervention over surgery in the above mentioned defects.



No chest scars
Cosmetic advantage
Shorter hospital stay
Day care procedure
No blood transfusions
No heart lung machine
No prolonged ventilation

He further talked about Hybrid Procedure wherein both, the cardiologist and the surgeon join together to perform the correction. (for.eg) A baby with Apical membranous VSD. Here, the procedure undertaken is as follows: Once the chest wall and the pericardium is cut open under the guidance of an *echocardiographic probe* placed on the surface of the right ventricle and with the help of the epicardial echo, a needle is passed directly into the anterior wall of the right ventricle and as the defect is identified, a guidewire is advanced through the VSD and the needle is removed and a introducer sheath is passed and through which the device is introduced and the defect is closed. Throughout the procedure the cardiac activity is maintained and the recovery after VSD closure is achieved very fast. The surgeon and the cardiologist work together here.

Other corrective procedures in Acyanotic heart diseases are
1) pulmonary valvotomy, 2) aortic valvotomy
3) coarctation stenting, 4) PA stenosis stenting

Pulmonary and aortic valvotomy are similar to coronary angioplasty wherein coronary balloon is used (Such procedure is used mainly in Aortic stenosis). In critically ill neonates with significant heart failure, renal failure and duct dependant systemic circulation, the intervention helps in instantaneous improvement in ventricular function.

Neonatal Balloon Aortic Valvotomy is a challenging situation. It involves difficulties in a) vascular access, b) hardware issues.

Vascular access: is when the baby is in shock, the blood vessels will be collapsed wherein a surgeon will be needed to perform a cut down in the carotid / axillary artery to identify the blood vessel. Some times blind puncture guided by anatomic landmarks shown by fluoroscopy in groin is done to cannulate the femoral artery.

Hardware issues are all the devices should be miniatured to suit the newborn.

In Coarctation - Stenting is the standard intervention. In Native coarctation (coarctation treated for the first time) for a child at the age of six and above - stenting is performed. The stents used are dilatable in future to adult size.

In small and sick children with multi organ failure and with severe pulmonary hypertension the cardiologists will resort to balloon dilatation a less morbid procedure. The common complication in coarctation stenting is after the procedure,the ductal tissue in the region of coarctation entering into the pulmonary artery gets progressively cicatrized and makes the coarctation to recur within 3 - 6 months. But then it may be easy to perform surgical intervention once the child is grown by 6 months.

Intervention in cyanotic heart disease:
Spectrum of interventions are

(1) As Initial palliation; (2) As Curative treatment;
(3) As adjuncts to surgery.

In **(1) Initial palliation,** it includes a) Atrial septostomy, b) Stenting of the patent arterial duct,

C) RVOT balloon dilation in Tetrology of fallot's.

(A)Balloon Atrial Septostomy: This is the first palliative treatment to come in 1960's.It was not considered as an intervention then because surgery was needed later for the correction of Transposition of arteries.

In transposition of great arteries - There is parallel circulation-meaning (venous blood circulates from right atrium >right ventricle >Aorta and the pulmonary circulation gets only oxygenated blood) when a hole (i.e) ***septostomy is created*** then the saturated blood is allowed to pass from left atrium into right atrium and venous blood from right atrium into left atrium.

Other reasons for septostomy are: Tricuspid atresia / stenosis, Mitral atresia / stenosis, HLHS, TAPVC with restricted PFO, Univentricular heart with poor inter atrial communication.

The procedure is as follows: An inflated balloon is pulled back against foraman ovale to create a tear. This is done by using femoral vein or umbilical vein, with the help of an echo guidance,and is performed in ICUor with a Fluroscopy in cath lab. In classical TGA-PFO is easily torn. In Non TGA condition Balloon septostomy will not work because of restricted PFO wherein **Static balloon dilation** is used.

In situations were the atrial septum is very thick, **Blade septostomy** is employed. Blades are available in three different sizes from 10-20mm. In certain conditions, like in HLHS wherein when the atrial septum is dilated, it collapses again and in such situation atrial septal Stenting is performed.

(b) Ductal stenting; {Second palliative procedure}

In Neonatal period, when there is pulmonary atresia with duct dependant pulmonary circulation - the patency is maintained by using Prostaglandin E1 infusion. This is maintained for a period of 7-10days.

Within this period, stenting is performed. In the past, BT shunt was performed. But since that procedure has substantial mortality and complications (lung collapse, lung infection, septicaemia) during neonatal period, Ductal stenting is opted for. In this procedure, a catheter is passed through ductus arteriosus and a wire is passed through and then a stent is put across. The correction achieved is equivalent to BT shunt.

In duct dependant systemic circulation{HLHS}large stent measuring 8mm is used.

C) Balloon dilation in Tetrology of Fallot:

Surgeons are comfortable in complete correction of TOF by 3 months. Balloon dilation is mainly useful in bridging the time between neonatal period and the time when surgery could be performed.

2) Curative Therapy:It includes

a) Balloon pulmonary valvotomy in critical Pulmonary stenosis in newborn
b) Perforation of the pulmonary atresia with intact ventricular septum
c) Closure of pulmonary AV fistula coil closure

In critical pulmonary stenosis in newborn - where the blood passes from right atrium through the patent foramen ovale into left atrium, is associated with critical cyanosis. In this condition, when balloon valvotomy is performed antegrade blood flow from right ventricle into pulmonary artery is restored and this results in total cure.

In AV fistula - It is an abnormal connection between pulmonary artery and pulmonary vein associated with intense cyanosis.{when this condition is associated with osler weber rendu syndrome / heriditary hemorrhagic telangiectasia it is diffuse} In localised Avfistula, catheter closure is feasable and curative.

In pulmonary atresia with intact ventricular septum - the atretic membrane is perforated with a guide wire or radiofrequency wires and a balloon is passed through and opened up thus a complete cure is obtained.

3) **Interventions as Adjuncts to surgery:** Here the following conditions were explained *a)Pulmonary artery stenting* - when the artery gets narrowed due to fibrosis relieved by performing stenting of the Pulmonary artery(PA).

b) Stenting of RV to PA conduits

c) Recanalization of blocked BT shunts - when the block is due to thrombosis or diffuse fibro myo intimal proliferation within the shunt - a wire is passed through the block and dilatation is achieved
d) Coil closure of Aortopulmonary collaterals
e) Closure of Aorto pulmonary shunts
f) Closure of additional muscular ventricular septal defects - when TOF is surgically treated
g) Coil occlusion of venous collaterals - after bidirectional shunt or Kawashima shunt surgery for cyanotic heart disease
h) Pulmonary valve implantation

Catheter Interventions for Cyanotic Heart Disease

- In many acyanotic conditions, interventions may be the only definitive treatment.
- In cyanotic conditions, certain interventions are performed in the following situations
- As initial palliation
- As curative treatment
- As adjuncts to surgery

Interventions for Congenital Heart Disease: Conclusions

- Interventions for congenital heart diseases are a cooperative effort of pediatric cardiologists and surgeons to provide the best possible care
- In about 40%, it is possible to effect a effect a change by interventions alone

Discussion: When Dr.Sujatha raised a question about *the device which is deployed in early infancy or early childhood - whether it has to be replaced as the child grows,* Dr. Sivakumar explained that the Device once deployed (which is madeup of nitinol) gets endothelialised within 6months and it becomes a part of atrial or ventricular septum and the remaining portion of the septum grows while the device remains the same.

But in Stenting once the stent diameter is inadequate for the age and weight of the child, these stents can be stretched by **dilation** which can be performed as the child grows to adult size. Usually the growth after 16yrs is negligible so around 16yrs, final dilation is performed.

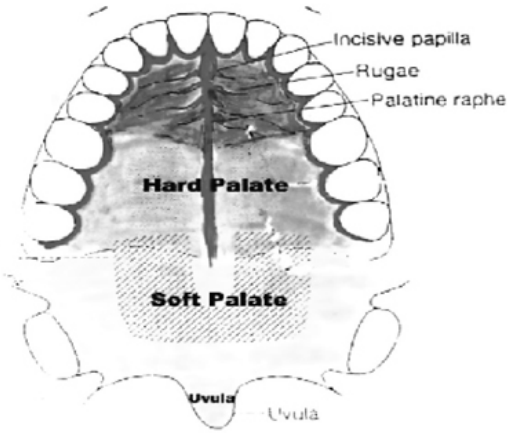
He also spoke about the research which is going on-on **Biodegradable Stents**. These biodegradable stents are made up of **magnesium.** Magnesium, usually gets absorbed within 3-6 months and within 1 yrs there would be no material found. But in some cases magnesium gets absorbed with in 4weeks wherein the purpose of stenting willnot be achieved. **About Prophylaxis** for infective endocarditis - Dr.Sivakumar explained that Antibiotics and aspirin were used for a period of 6months and after the procedure is performed, no life long medication is needed.

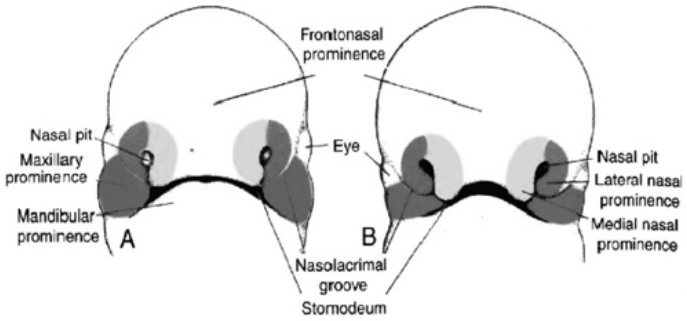
The Fourth & Final BDRI meet for this year started off with the welcome speech by our Director Prof.S.Suresh.

He introduced our special invitee Dr.Jayaraman.V an eminent plastic surgeon. *The main topic of discussion in this meet is about the Management of Cleft lip & Palate.* He also spoke about early detection of cleft lip and palate in ultrasound and how it fecilitates in counseling the parents, and about its correctability in isolated cleft defects.

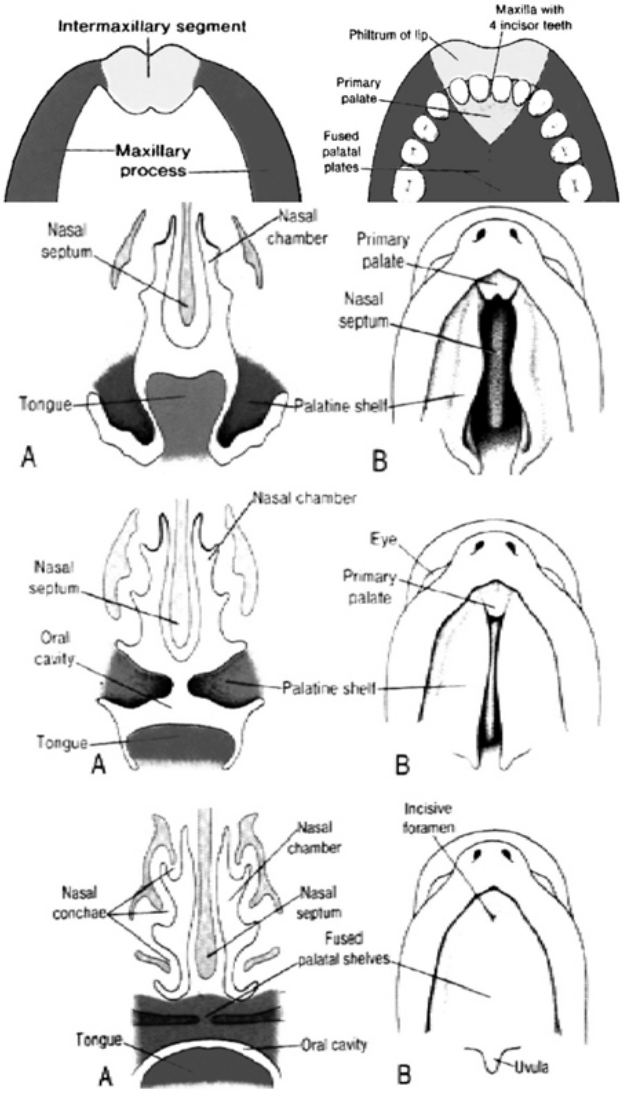
Dr.Sudha Ramakrishnan, ***Fellow in Fetal Medicine, MediScan*** spoke about facial clefts.

Her talk focused about the normal development of the facial structures and entailed the causes of cleft lip and palate and the detection of the same in ultrasonogram. **The cleft lip and palate-** Orofacial cleft lip is quiet a common congenital anomaly and occurs 1in 1000 live births. **The palate** is a tissue that interposes between the oral and nasal cavities. It has a hard palate and a soft palate. It has two parts. The **primary** and the **secondary palate** demarketed by the incisive foramen.





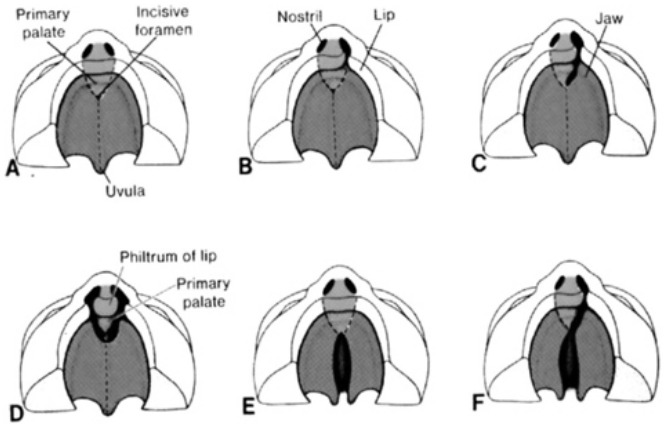
The fetal face starts forming as early as in the **fourth week** of the gestation. Palate starts forming within **five to eight weeks of gestation.** The process begins with the formation of the prominences. ***The fronto nasal prominence,nasal prominences (two in number), maxillary prominences (two in number), and a mandibular prominence.*** A groove is initially formed between lateral nasal prominence and maxillary prominence-known as **nasolacrimal groove,** which then develops into naso- lacrimal duct.To begin with all the prominences are laterally placed. Once the nasolacrimal duct is formed the lateral nasal prominence and the maxillary prominence begin to fuse and along with these, the median nasal prominence and the maxillary prominence also come close to each other. The fusion takes place in both in the superficial and deep aspects. Once the maxillary prominences fuse with each other, the middle segment which is called as **the inter maxillary segment** or the median palantine process gives rise to the formation of the **primary palate** which includes the philtrum of the upper lip, incisive foramen and the four incisor teeth. The lateral palantine process which also originates from the maxillary prominence, leads to the formation of the **secondary palate.** The area anterior to the incisive foramen is the primary palate and posterior to the incisive foramen is the secondary palate.



The primary palate is formed by the fifth week of gestation and the secondary palate formed by seventh week of gestation. The lateral palantine process is initially obliquely placed and the tongue is in between and placed high.When the lateral palantine process becomes horizontal, the tongue has to fall down and help in the fusion of the prominences.

The lateral palantine processes fuses along the midline, anterior to posterior - with themselves and with the primary palate. Thus the complete fusion of the maxillary process takes place. During this process when the tongue does not fall down, the fusion is affected and may lead to CL/CP . Eventually the nasal septum also fuses with the posterior part of the maxilla.

The secondary palate which is formed consists of two parts, a)A bony componant b)soft componant along with uvula. (Facial clefts arises when there is an error in fusion of the prominences at any level). Clefts-anterior to the incisive foramen are the anterior clefts and posterior to incisive foramen are the posterior clefts.The anterior clefts starts from the lip and extends behind while the posterior clefts arises posteriorly and extends anteriorly.



Clefts can be Unilateral, Bilateral, Central, Anterior-involving the lip - with or without involving the primary palate, Posterior-involving secondary palate with or without involving lip and primary palate or sometimes involving uvula alone.

Antenatal risk factors

- Parental history or sibling history of cleftlip or palate
- Sex : males are twice likely to have cleft lip and females are twice likely to have cleft palate.
- Race : common among asians and american indians.
- Drugs -phenytoin,benzodiazepines,sodium valproate and corticosteroids, folic acid deficiency.
- Maternal obesity
- Smoking and alcohol consumption > 10 units per month.

Associated syndromes

- Apert's
- Goldenhar syndrome
- Di George's syndrome
- CHARGE
- Pierre Robin sequence
- Trisomy 18

Ultrasound diagnosis of Cleft lip and Palate

Fetal face can be viewed in three planes in ultrasonogram to detect any deformity. Our Unit protocol at Mediscan is to view the fetal face atleast in two of these planes on a routine basis at target scan. The planes are as follows:

1)Coronal plane: This helps in viewing the fetal nose,lips and premaxillary triangle. The premaxillary triangle is formed by the premaxilla/primary palate as the base, arms of the triangle is formed by the frontal processes of the maxilla and they meet together in midline with the nasal bone.The integrity of the triangle rules out cleft of the primary palate.Deformity in secondary palate cannot be identified in this plane.

This protocol was initially proposed by Prof.S.Suresh et al (MediScan) in 2002 and was published in 2006. This was later on adopted as Retronasal triangle in 2009 by Sepulweda et al.

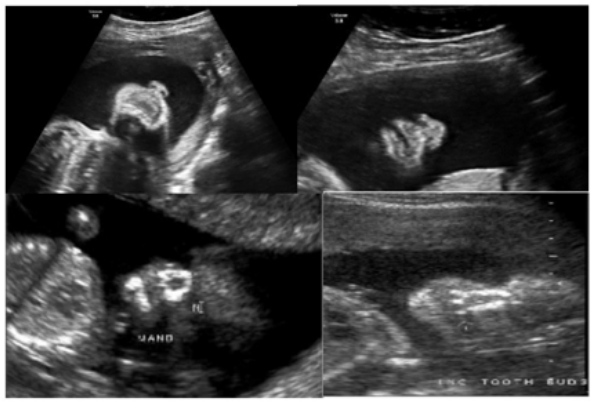
2)Axial plane: This plane allows visualisation of the nose and the alveolus with the tooth buds of the incisors and the palate. This plane gives a better assessment of the extent of the cleft palate.

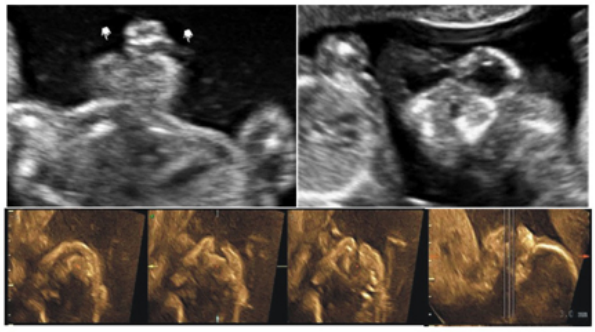
3)Sagittal plane: The nasal bone, premaxilla / primary palate, secondary palate and the soft tissues in the face can be imaged in this plane. A discontinuity in the palate, leads to the suspicion of cleft palate if the sagitttal view is on the same side of the defect. Soft tissue protruberance in this plane may give a clue to bilateral clefts.

Isolated clefts in secondary palate are difficult to be diagnosed on an antenatal ultasound

Ultrasound assessment of lip and palate

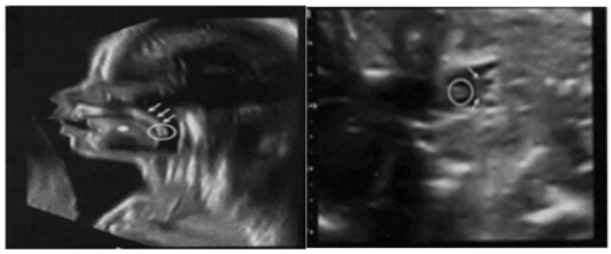
- Soft tissue in the upper lip
- Pre-maxillary triangle





2D imaging helps in detecting defects in primary palate and when defects of the secondary palate are suspected , 3D imaging plays an important role in establishing the extent. Viewing defects in secondary palate on 2D is difficult, because of the position of palate deep inside the face, shadows from the facial bones and the presence of soft tissues like the tongue may interfere with the imaging.

Additional signs to evaluate cleft lip and palate



When the fetal face is viewed in the sagittal plane,uvula appears as an (equal 2)"=" sign in the posterior part of the palate and in the transverse view, the uvula appears as an (equal2)"=" sign in front of epiglottis.

The presence of"=" signs indicates that the uvula is intact and is an indirect clue that the secondary palate may be intact. When the sign is not viewed, then 3D imaging is opted to confirm the diagnosis. All the above mentioned views can be viewed as early as 11-14 wks. We routinely image the face in the sagittal and coronal views in the first trimester ultrasound.

Isolated clefts have a good prognosis. Sometime clefts may be associated with chromosomal disorders. In suspected cases Direct testing is offered to ruleout associated anomalies and parents should be counseled in detail as in Multidisciplinary protocol,

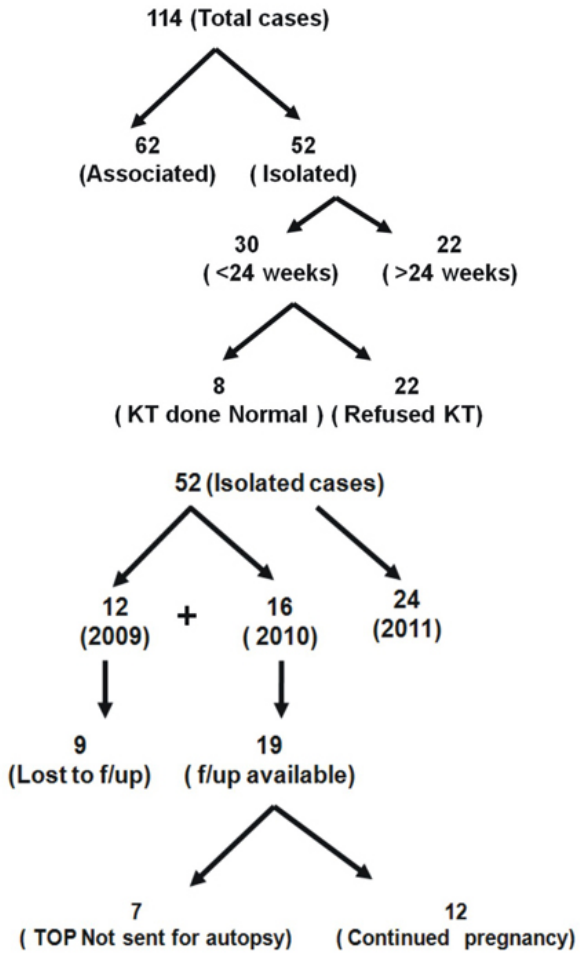
Multidisciplinary team

- Genetic counselor
- Pediatrician
- Plastic surgeon
- Pediatric dentist
- Otolaryngologist
- Audiologist
- Speech pathologist
- Social worker

Dr.Sudha spoke about the support group - SMILE TRAIN - a non profit organisation started in 1999 with it's headquarters in Newyork which does a lot of corrective procedures free of cost. Around 600,000 surgeries has been performed worldwide and over 300,000 surgeries in India alone. A documentary film on the defect was taken in India ***Smile Pinki.***

Statistical Data of Cleft lip and Palate by
Dr.Sheethal, Fellow, MediScan

Dr.Sheethal talked about the data collected for the period **Jan 2009 - Nov 2011.** Total number of cases evaluated were 114. For the year **2009 -** 39cases, **2010 - 36cases,** upto **Nov 2011 - 39cases.** In the **114 cases,** 62 were with associated anomaly, and 52 were isolated clefts. For this year,percentage of isolated clefts were in more number. Among the Isolated clefts,58% were left sided clefts (common), right sided 31% and Bilateral clefts were 11%.In the case study, 52 cases -which were isolated defects were evaluated as follows.



In the 30 cases within 24weeks of gestation,when direct testing was offered,only 8 of them accepted, and for all 8 the KT was normal.

In the **follow up** case study for 2009 - 2010
Total cases - **28.**

9 cases - had change of address,
In remaining **19cases**
(a)**7 -** opted for **MTP**(not given for autopsy)
(b)**12** - Delivered.In these 12 cases **apart from 2** all other cases were **FTND.**

Out of 12 patients, 5 had come to MediScan for FTS and in 4 of them the anomaly was detected and in 1 patient - it was detected in second trimester scans**.**

Follow up of 12 cases

- Sex: 3 females, 9 males (1:3)
- Age: 1 + 2/10 to 2 +2/10 (Median 2+6/12)
- Birth weight: 1.1 kg to 3.5 kg (2 LBW were preterm)
- 9 out of the 12 babies had 2 stage surgeries
- 3 had only 1 surgery
- 1 is advised 3rd stage of surgery
- Timing for first surgery was 3 - 4 months
- Timing for second surgery was 10 -12 months

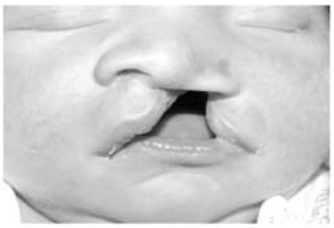
Dr.Jayaraman ***M.S(Gen.sur),Mch (plastic), Dip NB (plastic) Ph.D (plastic), FICS, Professor & Head Department of Burns, Plastic and Reconstructive Surgery, Kilpauk Medical College, Chennai,***

Dr.Jayaraman started his presentation with statistical study from Kilpauk medical college.

Types of Clefts

Nature and type of defect	Side of the defect	No.of Patients	%
Cleft Lip Group I	Right	186	5.69
	Left	309	9.45
	Bilateral	95	2.9
Cleft lip Group 1a	Right	249	7.61
	Left	319	9.76
	Bilateral	11	0.34
Group II Cleft Palate		533	16.3
Cleft lip and palate Group III	Right	608	18.60
Left	423	12.94	
	Bilateral	517	15.82

The study explained the nature and types of clefts and in that the left sided cleft lip was most common. He further elaborated on the significant incidence of cleft lip and palate in consanguinous marriages. In Total 26.52% were affected in consanguinity wherein. In I degree -17.2% and in II degree - 9.28% were noted. He was happy to inform that there was a reduction in consanguinous marriages in the statistical data for the period of ten years (1992-2002) which was probably due to effective cleft lip and palate awareness programme and there was decrease in medical termination of pregnancies and in usage of drugs in early pregnancy (First Trimester) after that programme.

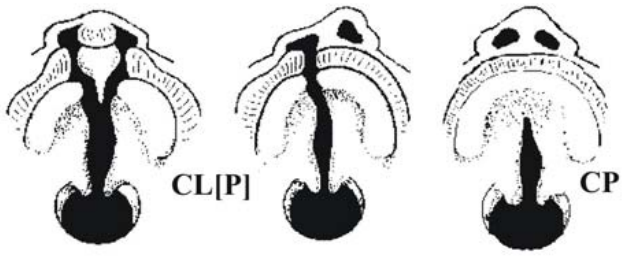


Types of Clefts

Oro-facial clefts include the cleft of the lip with or without cleft palate (CL[P]) and the isolated cleft palate (CP)

Classification

- Unilateral / Bilateral
- Complete / Incomplete
- Cleft palate / complete / incomplete
- Isolated / syndromic
- Cleft lip and palate



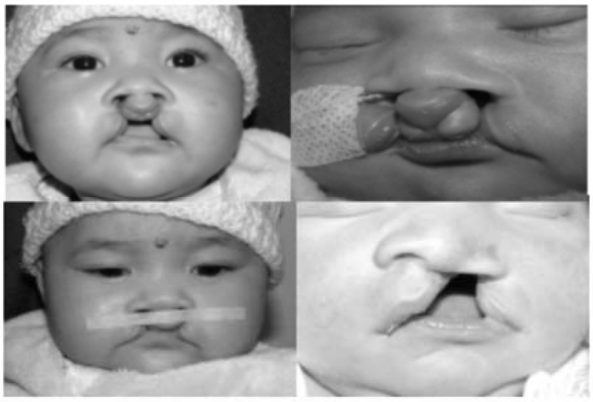
Symptoms

- Failure to gain weight
- Nasal regurgitation when bottle feeding
- Poor speech
- Misaligned teeth
- Growth retardation

Risks in clefts

- Ear infections
- Hearing loss
- Dental cavities
- Displaced teeth

Initially when repair of cleft lip and palate was started in India at Nagpur, cleft lip was tackled first and the cleft palate was corrected as second step.In recent times, for more of aesthetic value,correction of cleft lip is undertaken within six months of age and cleft palate repair is undertaken between 10months - 18 months of age. Dr.Jayaraman was quoting his observations during his training period in England were he had seen Intra uterine repair of clefts being undertaken. He interpreted that after such procedures, as the child grows, because of the difference in the growth at the operated site and normal area-the deformity persists to a noticeable extent. Ideally the repair is done within 2years of age. In bilateral clefts, the distance inbetween the clefts is reduced by plastering and keeping the line of joint close to midline and acheiving good results in corrective procedures.



Major problems faced by the children with cleft deformity are difficulty in suckling, swallowing speech, hearing and malocclusion.

Treatment: The first stage of treatment begins at 3-9 of age. More than one surgery is needed only when there is extensive damage. As the initial repair is performed, in some cases, nasal repair is also done but as the nasal cartilage grows with age the possibility of the deformity showing up again arises. Long list of procedures are required for these children.

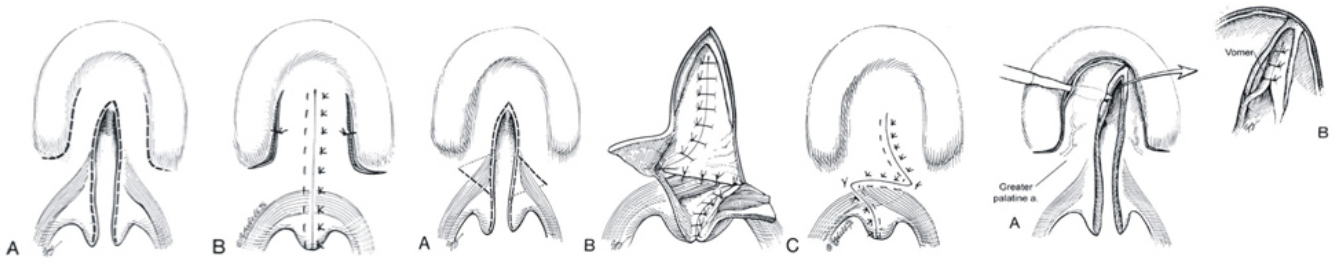
Long list of list of procedures these children undergo

- Neonatal orthopaedics-orthodontist(take care of the palate repair)
- 3-12 months repair of lip and anterior maxilla - *plastic surgeon*
- 9-18 months repair palate-plastic surgeon
- 5 yrs revision of lip repair-plastic surgeon
- Grommets placed due to 'glue ear'- ENT
- 7-10 yrs orthodontics/ ABG / repair OAF- oral surgeon
- 12-18 yrs orthodontics/orthognathic surgery

Surgical aims in cleft lip repair

- Reposition ala
- Restore nasal floor (when nasal floor is not repaired fistula can develop between mouth and nasal cavity)
- Lengthen columella on cleft side to equalise-philtrum is formed better
- Lengthen medial lip segment (typically, lateral lip segment has enough length)
- Reconstitute symmetrical vermillion roll (muscle repair is important)
- Restore dry vermillion medially (typically, lateral segment has enough dry vermillion)
- Align wet vermillion to dry vermillion line ("wet to dry line"
- Realign and correct abnormal insertion of orbicularis oris muscle
- Reconstitute philtral column (typical by placing the scar at the philtral column site)

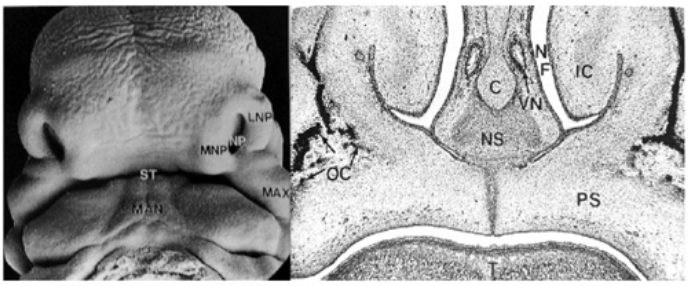
The surgical procedure is performed in such a way that the uvula is restored and inner and outer muscular layers are sutured methodically

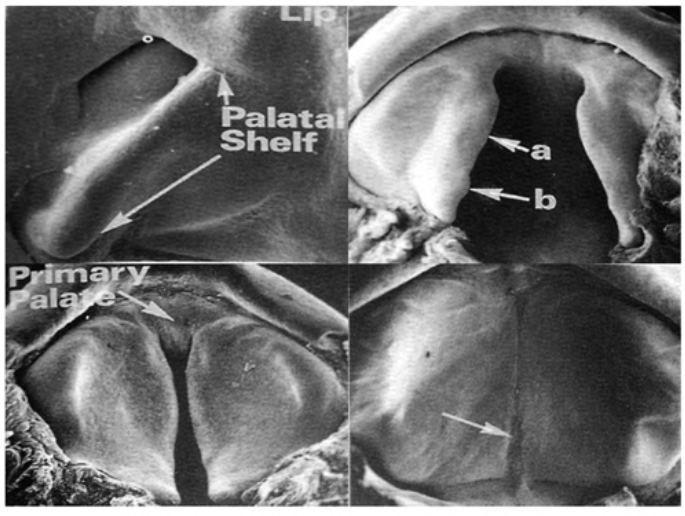


Following Dr.Jayaraman,

Dr.Harikrishnan-*BDS, MDS, FDSRCS(Eng), FCROR(Taiwan),FPFA, MBA(HM), PGDMLE*

Dept of Orthodontics - also spoke about the commonst form of orofacial clefts - cleft lip and palate wherein the orthodontists have major contribution in correcting the defects. He initially briefed about the pathogenesis of cleft lip and palate

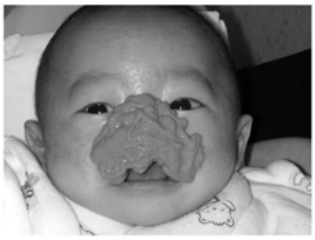




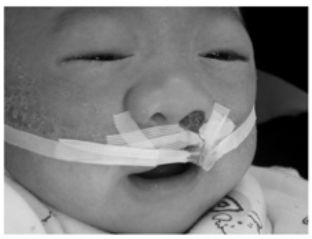
Treatment protocol (Universal):-The protocol is common for orthodontists and plastic surgeons.

- At birth - paediatric full examination, feeding advice, treatment plan, counseling
- Day 2 - orthodontic feeding plate, NAM appliance, taping for Bilateral CL.
- 3 months - CL repair, ENT exam
- 5 yrs - fascial assessment done by orthodontists and maxillary hypoplasia is corrected
- 7 yrs - preventive and interceptive orthodontics (repair the excess mandibular growth)
- 9 -11 yrs - Pre alveolar grafting and leveling up of teeth is done by orthodontists and alveolar bone grafting is done by plastic surgeons. Nowadays these procedures are done within 10yrs before the growth of the cannine teeth.
- 12 - 15yrs As the permanent teeth are formed, the non alignment is corrected. (tertiary orthodontic care)
- 16 yrs - Maxillary hypoplasia is taken care off-by
- Advancement osteotomy and pushback osteotomy (orthognathic surgery)
- 18 yrs - rhinoplasty, prosthetic rehabilitation
- 20 yrs - counseling is done (genetic counseling)

NAM - impression

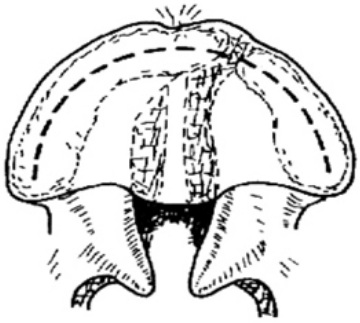


Strapping to maintain uniformity

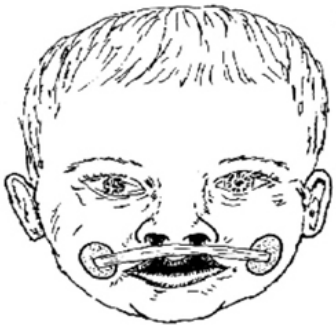


Arch Aysmmetry

Arch Alignment



Premaxilla taping



Arch Alignment

Dr. Harikrishnan concluded his presentation with this note - Managing cleft lip and palate is a team work wherein lots of effort is put in by doctors as well as the co-operation by the parents which goes hand in hand to acheive the objective.

Nurse Girija- working at Child Trust Hospital for 15yrs - explained the feeding methods for the baby with cleft lip and palate.

a)New born: To be fed with syringe / paladai / spoon **b)** Should not be allowed to suck directly. **c)**When deformity is severe, Ryles tube feeding is advised **d)** While feeding the baby - the Head end to be raised and to be fed slowly

Discussion

The queries discussed at the end of the presentation were;

1.When the child is brought for correction of the defect after 2yrs - Dr.Jayaraman explained that the normal protocol is to do the correction from 3 mths, when the child is brought at a later age, the correction is performed methodically along with the lengthening of uvala.The presence of nasal tone is the main sequlae which is dealt with proper exercise to the larynx-like pronouncing the specific syllables continuously as per advise.The exercise is to be practised by all operated /corrected babies.

2.Dr.Jayaraman assured that the operative procedures have improved in recent years and with the joint involvement of the multi-disciplinary team,the outcome is very fruitful and when the deformity is identified antenatally a clear cut counceling is helpful in guiding the parents.

3. Speech therapy - Post surgically for CL/CP patients to prevent nasal tone and absence of clarity of speech, appropriate speech therapy has to be insisted early and rigourously.

4. Discussion continued about the option of Medical termination of pregnancy. CL/CP when diagnosed antenatally in majority of cases MTP is opted only when the condition is associated with other anomalies. Isolated CL/CPs are rarely terminated.

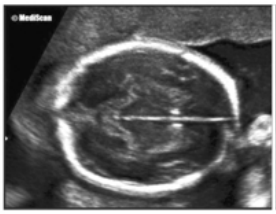
5. Corrective procedures are well advanced in recent years but, cost and continuous followup are to be considered.

The final BDRI meet for the year 2011 concluded with Prof.Suresh acknowledging and thanking our chief guest and participating members.

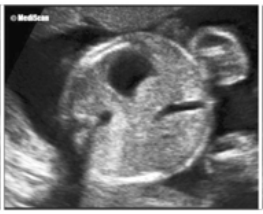
FETAL CARE RESEARCH FOUNDATION

18 - 20 wks scan Standard images for Rule of Three

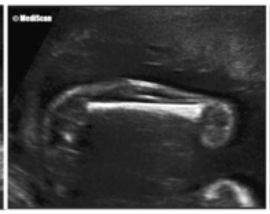
MediScan
Ultrasound | Fetal Care | Genetics



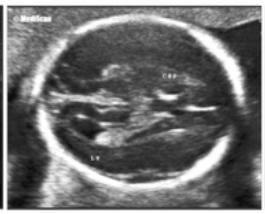
Biparietal Diam. (BPD) / HC



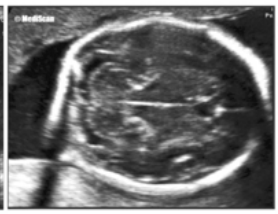
Abdominal Circum. (AC) (Stomach)



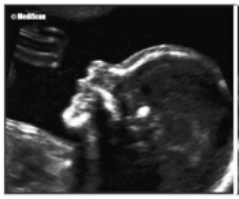
Femur Length



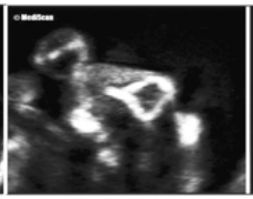
Lateral Ventricle & CSP



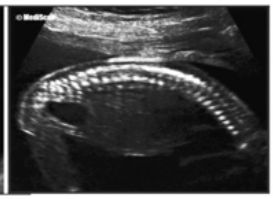
Cerebellum / Cisterna magna



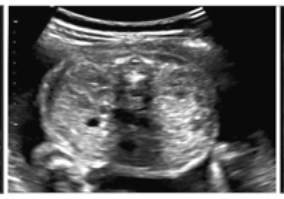
Face - Profile



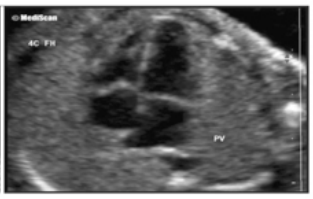
Premaxillary Triangle



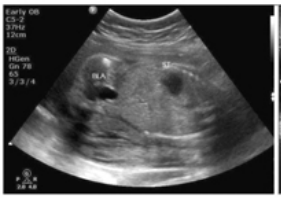
Spine



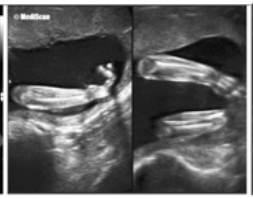
Transverse Spine & Kidney



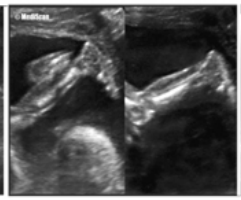
Four Chamber Heart



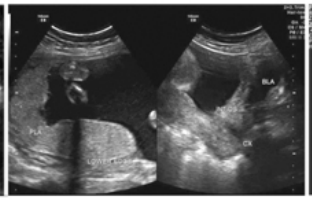
Bladder



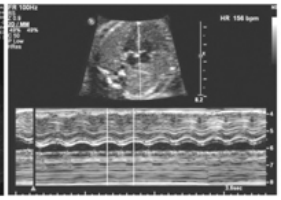
Upper Limbs



Lower Limbs lateral



Placenta + Cervix



M Mode

ENTIRE BDRI TEAM WISHES AII THE MEMBERS

"A HAPPY AND PROSPEROUS NEW YEAR"

Support a National cause...
Join the league...
It is just a click away!!!

Browse our website: www.mediscansystems.org and click on the Registration form. You will be enrolled in BDRI and guided further.

For queries please contact

BDRI Co-ordinator
Call: +91 - 44 - 2466 3141/ 98403 29385
email:bdri@mediscan.org.in

This news letter is available at http://www.mediscansystems.org. quarterly - January, April, July and October.
Published by Fetal Care Research Foundation, 197, Dr. Natesan Road, Mylapore. Chennai - 600 004.
For Private circulation Printed at "The Print Shoppe" (Print Supplies), Ayanavaram, Chennai - 600 023.