B D R News

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

Volume 4 Issue 4: October 2003

PROCEEDINGS OF THE BIRTH DEFECTS REGISTRY MEETING HELD ON 31st OCTOBER 2003.

The last meeting of the year was held on 31st October 2003 at Mediscan Systems Annexe, Chennai. Dr. S. Suresh welcomed the gathering. The topics for the evening were "Embryology and USG Imaging of the Fetal Spine" and "Management of Congenital Spinal Disorders". Dr. Suresh announced the news about the inauguration of two new registries recently commissioned at Lalgudi and Trichy. He also appealed to Chennai members to motivate others and enhance the memberships in Chennai. Dr. Sujatha Jagadeesh introduced the guest speaker Dr. K. Sriram, Consultant Orthopaedic Surgeon with special interest on spinal deformities in children. The session concluded with vote of thanks proposed by Dr. Suchitra Ravishankar. Given below are the excerpts of the presentations.

EMBRYOLOGY AND USG IMAGING OF THE FETAL SPINE

Dr.Indrani Suresh, Director, Mediscan Systems

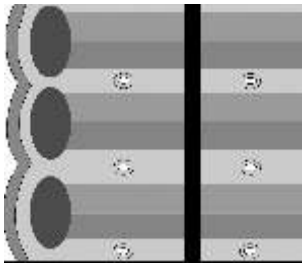
Embryology

The development of fetal spine starts from the third post conceptional week and is completed by 8 weeks. The two important processes in spinal development are "segmentation" and "formation". Inductive influence of notochord and neural tube are essential for the development of vertebral column.

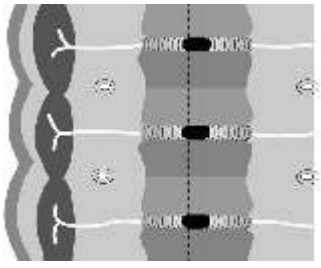
Segmentation Process:
Primary segmentation**:**

Intraembryonic mesoderm on either sides of midline condenses to form para axial mesoderm which gets segmented to form somites. Somite differentiates into *sclerotome, myotome* and *dermatome*. Sclerotome gives rise to vertebra, myotome to muscle and dermatome to dermis. Sclerotome of each somite has a cephalic and caudal portion Each segment has its nerve, and an artery. The artery is present in between the segments.

Primary segmentation



Resegmentation



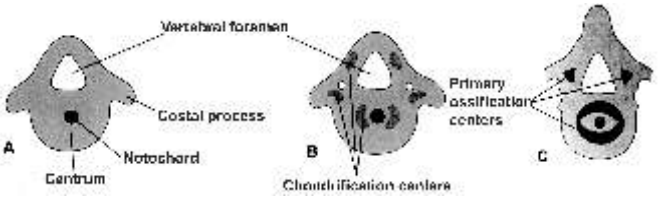
Secondary resegmentation

Resegmentation occurs by the union of caudal portion of one segment with cephalic portion of the inferior segment. This leads to positioning of nerve in the intervertebral space and artery in the middle of the segment. Notochord give rise to nucleus pulposus of the intervertebral disc and surrounding mesenchyme forms annulus fibroses.

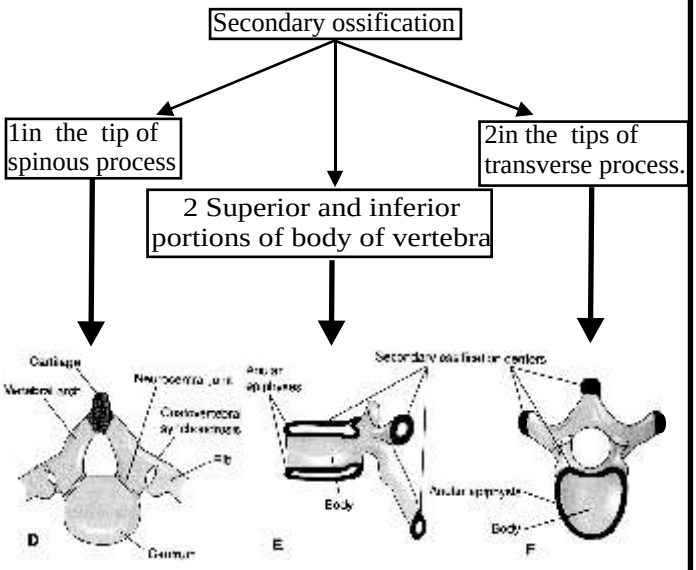
Ossification of vertebra:

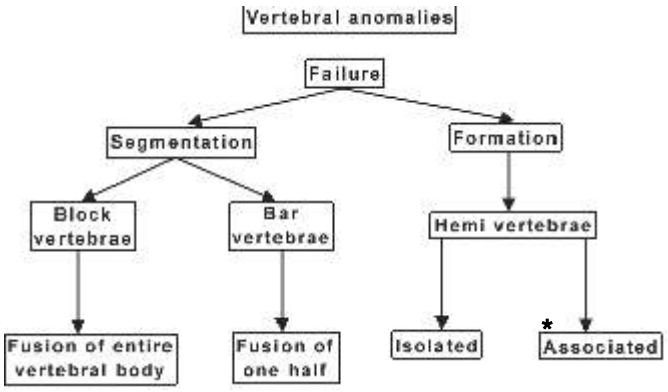
The process of occsification starts after the completion of vertebral cartilagenous module.

Primary ossification



There are three primary ossifictions centres - one anterior, two posterior, present in fetal life. Anterior ossification centres start its development at mid thoracic level which would progress in cranial and caudal direction simultaneously. Posterior ossification centres start at cervical end and progress in caudal direction. Secondary ossification takes place in post natal life.





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Anomalies associated :-

1.VACTERL 2.VATER 3.Jarcho Levin syndrome (spondylothoracic dysplasia) 4.Klippel Feil anomaly 5.Goldenhar syndrome 6.Neural tube defects - (Diastematomyelia, tethered cord, meningomyelocele)

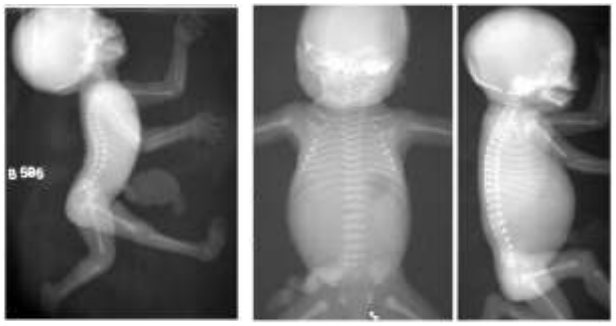
USG imaging of the fetal spine

Ultrasound is an excellent modality for imaging the fetal spine . We are familiar with X-ray appearance of spine in the antero-posterior(AP) and lateral(LAT) view. In AP view the vertebral body and pedicle are seen. In the lateral view the body and laminae are seen. Similarly, while imaging the spine in utero with USG, the anterior ossification centre represents the vertebral body and the posterior centre represents the vertebral arch. Imaging of the spine is done in three planes - coronal, sagittal and transverse. The normal fetal spine is "C" shaped & adult spine is "S" shaped(curvature).

Curvature of spine

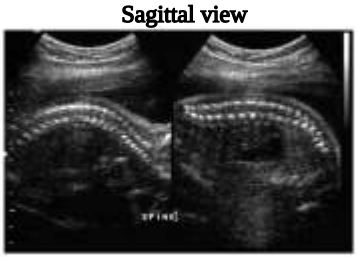
Abnormal Normal

Arthrogryposis

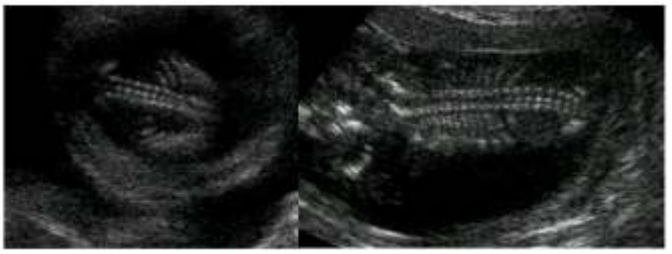


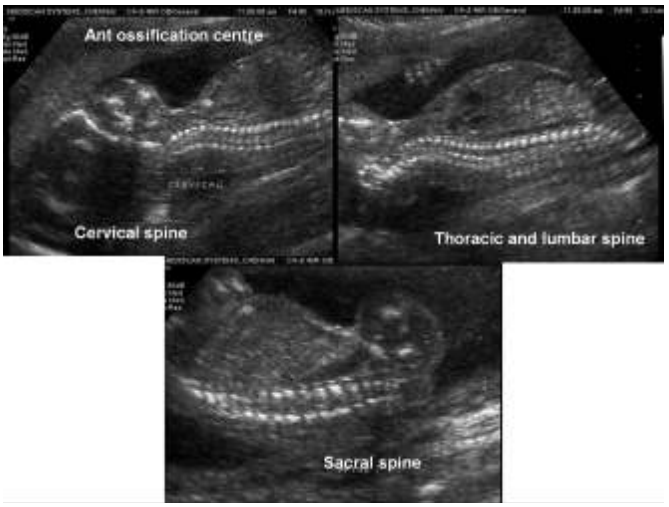
Imaging of Vertebral column

Sagittal view

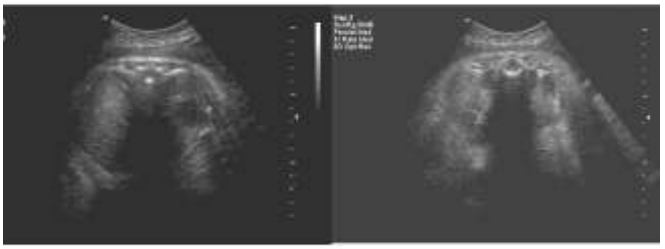


Coronal view





Transverse



Three dimensional imaging of the spine :

1.3D USG helps in obtaining a panoramic view of the thoracic cage.
2.Offline analysis of the data can be done later
3."Walk through" option in 3D increases the accuracy of diagnosing hemivertebra and block vertebra

Limitations in imaging

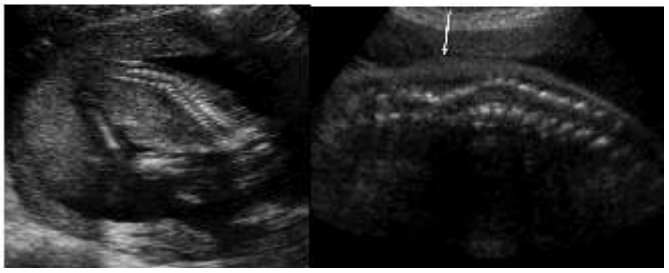
Posterior position of spine, Oligohydramnios and very early gestation less than 16 weeks or very late gestation more than 36 weeks are the major limitations of imaging the spine.

Etiology

Vertebral anomalies may be of multi factorial or single gene or unknown etiology. Vertebral defects may be isolated or associated with malformations like cardiac and genito urinary anomalies, neural tube defects, talipes equino varus and sacral agenesis. They can also be a part of syndromes such as Jarcho Levin, Klippel Feil syndrome. Spinal defects at thoracic level are associated with cardiac anomalies and defects at lumbar level have association with genito urinary problems. These associations are due to common embryological (mesenchymal) origin of structures involved.

When to suspect spinal anomaly?
Persistent abnormal curvature of the spine should raise a suspicion of a spinal anomaly, which warrants a meticulous search for the problem in all 3 planes. Abnormal curvature leading to kyphosis and scoliosis can be secondary to a large omphalocele or body stalk anomaly.

Hemivertebra
Persistent angulation of fetal spine - Longitudinal view



Counseling

Accurate prenatal diagnosis of vertebral anomaly and associated anomalies are very important for counseling the patient. Isolated vertebral defects have good prognosis. Even though associated anomalies can be suspected during prenatal imaging, some of them can be diagnosed only after birth.

Mediscan Data : 9 cases with isolated hemivertebrae were followed up over a period of four years at our centre. Of the 9 cases four children are growing well without any problems. One case was found to have multiple anomalies on autopsy. Followup was not available in four cases.

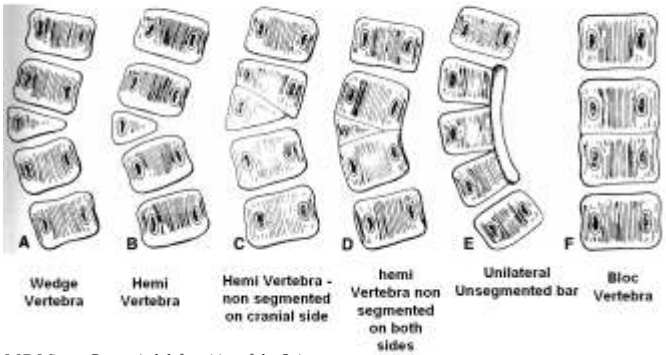
Dr. Indrani concluded her presentation saying that knowledge of embryology and anatomy of developing fetal spine facilitates better understanding of vertebral defects. A thorough search for associated anomalies and detailed postnatal clinical and radiological evaluation are essential for accurate diagnosis and management of spinal deformities.

MANAGEMENT OF CONGENITAL SPINAL DISORDERS

Dr. K. Sriram, Consultant - Orthopaedic Surgeon

Dr. Sriram's lecture started off with pictorial illustrations of various congenital anomalies of the vertebra.

Congenital Scoliosis - Classification



W.B.Winter, Congenital deformities of the Spine

These deformities alter the normal curvature of the spine. Scoliosis, for example, is progressive in 75% of cases. The unbalanced growth of the vertebra results in progressive deformity. The curve worsens in the first two years of life and during adolescent growth spurt.

Mc Master studied a large series of cases and found that the angulation became more severe if the vertebral anomaly was a unilateral unsegmented bar and contralateral hemivertebra at any spinal level i.e., lower / upper thoracic, thoroco lumbar and lumbar regions. In single hemivertebra or wedge vertebra the deterioration was much less and it was almost negligible in block vertebra. (See Table in page 4)

A systematic method of evaluating a patient with valuation of a patient with scoliosis should follow the steps mentioned below:

1) Complete physical evaluation
2) Complete neurological examination
3) Radiological evaluation
4) Accurate measurement and comparison of all X-ray films.

MRI is useful in the following situations:

- When there is a neurological deficit
- Clinical findings & X-ray are suspicious of neurological involvement
- Pre operative evaluation

Caution:
1) Entire spine should be imaged (not only the area of deformity)
2) Interpretation may be difficult in severe deformities.

Congenital scoliosis is **associated** with
Cardiac anomalies in 12%
Renal anomalies in 30%
Intraspinal anomalies in 38—41%

Management of scoliosis should have the following three objectives:

1) Early diagnosis
2) Anticipation of prognosis
3) Prevention of deterioration

The **follow up** protocol should include
A) Monitoring for progression by
- Clinical observation
- Photograph
- X-ray

B) Comparison of the initial, last and current films
C) Follow up throughout the growth period

If the curve is flexible, braces can be applied.

The **surgical indications** for corrections of congenital scoliosis are:

1) Severe deformity
2) Rapid progression of deformity
3) Small curves with bad prognosis

Median Yearly rate of deterioration (In degrees) without treatment for each type of single congenital scoliosis in each region of the spine (Mc Master)

Site of Curvature	Type of Congenital Anomaly					
	Block Vertebra	Wedge Vertebra	Hemivertebra		Unilateral Unsegmented Bar	Unilateral Unsegmented Bar and Contralateral Hemivertebrae
			Single	Double		
Upper thoracic	<1°-1°	★-2°	1°-2°	2°-2.5°	2°-4°	5°-6°
Lower thoracic	<1°-1°	2°-2°	2°-2.5°	2°-3°	5°-6.5°	5°-8°
Thoracolumbar	<1°-1°	1.5°-2°	2°-3.5°	5°-★	6°-9°	7°-14°
Lumbar	<1°-★	<1°-★	<1°-1°	★	>5°-★	★
Lumbosacral	★	★	<1°-1.5°	★	★	★

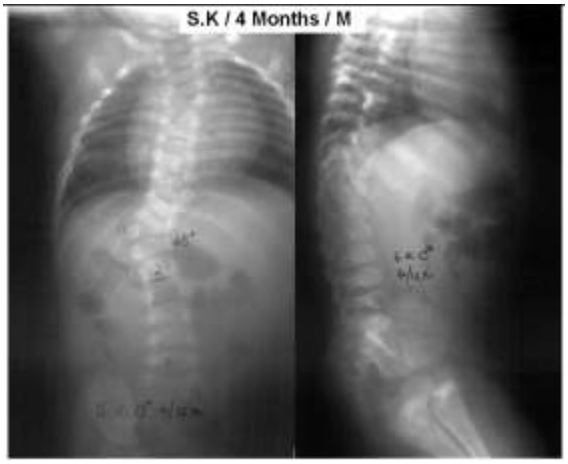
In these situations prophylactic fusion may be offered. Decision regarding type of surgery depends on age, site, type of anomaly and size of deformity.

Surgical Correction Methods:

1) Posterior spinal fusion is the gold standard. In unilateral bar, in situ fusion may be done. It is best done before 2 years of age. Fusion must extend to the entire curve or else progression of curve may occur.

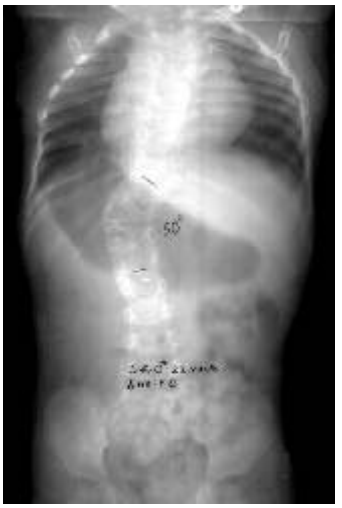
2) Combined anterior and posterior fusion allows greater flexibility of spine for further correction and eliminates growth potential.

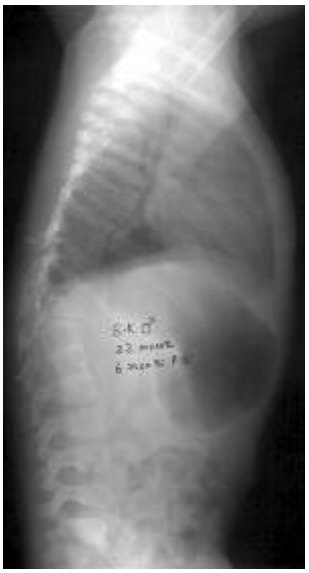
He illustrated the indications for surgical correction with excellent clinical and radiological photographs and how they were corrected.



Before correction:- Spinal deformity noticed at birth. Hemivertebrae at D8, D9 (L), D12 (L) D1 - L2 - 45° No deficit.

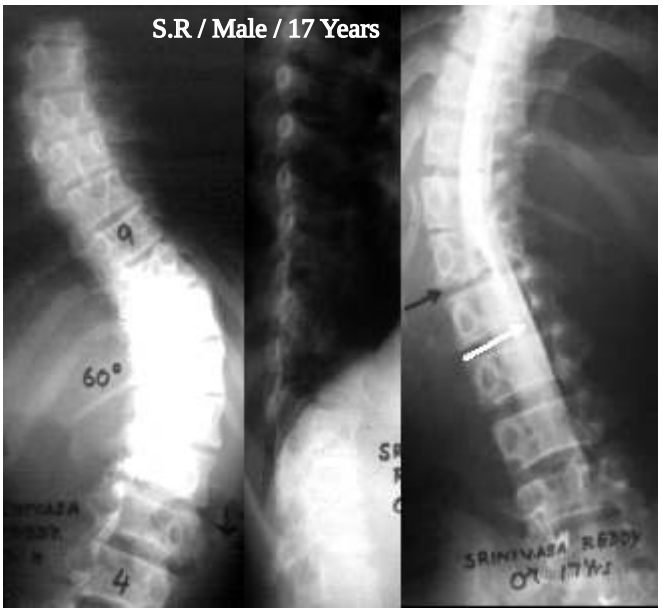
S.K / 22 Months / Male





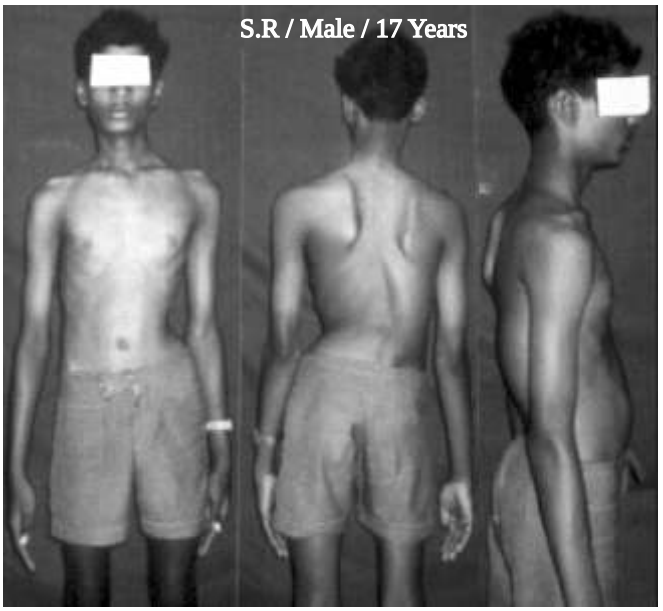
After correction:-

Anterior posterior fusion done D8-L2. Fusion is progressing well. Child is braced.

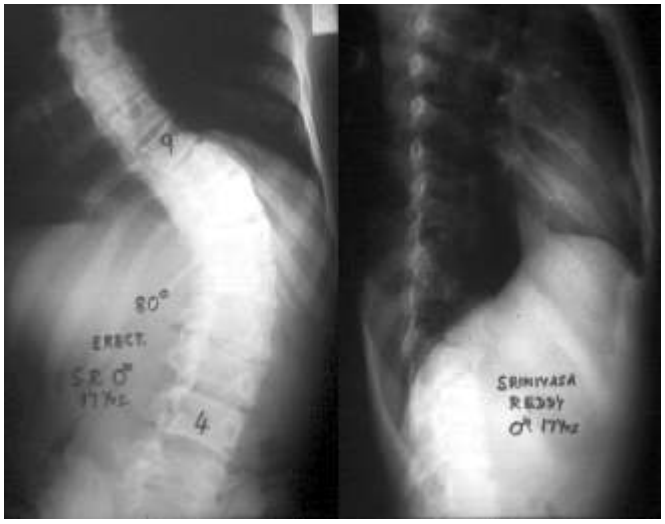


X-rays: Congenital scoliosis D9-L4 (60º). Imperfect segmentation at D12-L1.

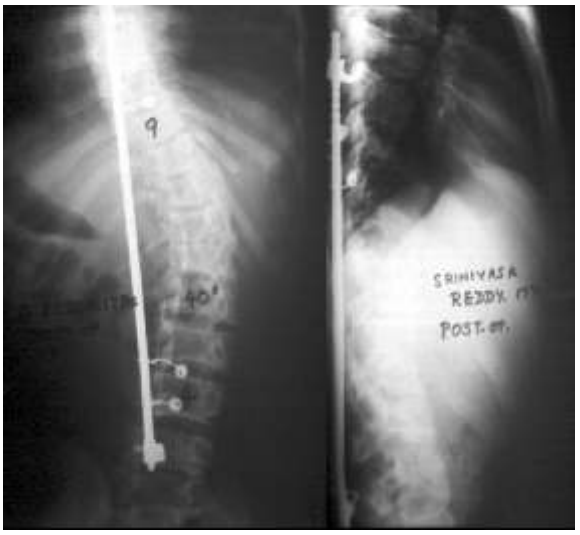
Myelogram- Diastematomyelia D12 - L1



Clinical appearance - S.R. Male 17 Years



Stage-1:
Excision of spur, Rapid progression of curve to 80°



Stage II: Anterior fusion D9-l3 and posterior fusion D7-l5 with instrumentation - spinal alignment satisfactory

The option of anterior and posterior convex epiphysiodesis is considered in patients with :-

1) Pure scoliosis 2) Good growth potential on concave side 3) Age below 5 years 4) Mild curves

In severe deformities—osteotomies, repair of psdeudarthrosis and extension of fusion are performed. Expected cosmetic results may not be achieved and risks are high.

He concluded that congenital scoliosis is a potentially serious condition and can lead to severe deformity.

Early diagnosis and anticipation are the keys to plan surgical corrections at appropriate time and lessen morbidity.During discussion the questions raised by the audience were:

1) *When to refer a child with hemivertebrae to an orthopaedic surgeon?*

At the earliest for initial assessment and 3 monthly follow up in the first year of life.

2) *Does spinal fusion reduce final height?*
No; but it should be noted that these children are genetically shorter than normal children.

Yet another milestone.....! Registries inaugurated

BDRI plaque received by Dr. Susheela Devi, Janet Nurshing Home, Nodal center Trichy BDR.

BDRI plaque received by Dr.R.Ramesh, Dr.Ramanathan Memorial Hospital, Nodal center Lalgudi, BDR.

Dr. R. Ramesh , Director, Dr. Ramanathan Memorial Hospital, Lalgudi and Dr. Susheela Devi, Director, Janet Nursing Home, Trichy have joined our band along with their colleagues by inaugurating BDR branches at Lalgudi and Trichy respectively. Both registries were inaugurated at a grand function held at Trichy on 26th October 2003 by Shri. Athi Krishnan , Joint Director of Health services, Trichy. Dr.Jayashree Ramesh, Dr. Victoria Johnston and Dr.P.R.Subramanian gave their felicitations. Both the nodal members were optimistic about making their respective centres as models for the others to follow.

BDRI has nominated Evita Fernandez Maternity Hospital, Hyderabad as a nodal centre to collect registry data from Hyderabad.

Due appreciations……

The central registry is very happy and thankful to our Erode BDR members and Salem BDR members for their meticulous furnishing of data. We do hope that our BDR family would soon grow by leaps and bounds and our dreams of building a national network of BDR come true.

BDRI WISHES ITS MEMBERS A VERY HAPPY NEW YEAR

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