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Editorial

Pediatric Education and Research
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Marching Ahead with Powerful Contingent of Adolescents -*the Demographic Dividends*- towards Renaissance of Primary Health Care

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An overarching theme to be observed in several of the primary health approaches that we have studied is the emphasis on comprehensive care. That is to say that in caring for people in all communities, social, economic, and historical factors should be considered together with health studies. It also means that the whole person is treated, not just the disease. At the Alma Ata Conference in 1978, among the contributors to this groundbreaking document was Dr. Carl Taylor. 30% of adolescents of our total 1.2 billion population eye eagerly towards Primary Health Care.

In one of the lectures delivered in Johns Hopkins University in 1987, Carl Taylor explained the roots of the Primary Health Care (PHC) and discussed the drafts by WHO and UNICEF leading to international conference on Health for all through PHC at Alma Ata in 1978. At this conference, medical, public health and global policy experts proclaimed: "The Conference strongly reaffirms that health is a state of complete physical, mental and social wellbeing, and not merely the absence of disease or infirmity[1]." In its boldness, the Declaration, asserts the Director-General of the WHO Dr. Margaret Chan, "articulated primary health care as a set of guiding values for health development, a set of principles for the organization of health services, and a range of approaches for a addressing priority needs and the fundamental determinants of health[2]." It also means that the whole person is treated, not just the disease- a holistic approach. Among the contributors to this groundbreaking document was Dr. Carl Taylor.

If we peep through the historical overview of the PHC by Carl, we observed that US clinicians first called PHC as the individual focus while community oriented primary care started with Kark in South Africa, then it went to US and Israel. Alma Ata label for PHC meant comprehensive (Horizontal) care but it became selective (Vertical) in 1984 due to the changing perception of donors, for example, the Bill Gates Foundation opted to focus on one global health issue-diseases specific and target to make maximum impact the eradication of Polio. In other words, health has moved from under-investment, to single disease focus, and now to increased funding and multiple new initiatives. Second well-known example of the selective primary health-care approach is the child survival revolution, championed by Jim Grant. This debate—between comprehensive and selective, horizontal and vertical, top-down and bottom-up—was the major topic of discussion in global health for the 1980s and 1990s, with few programs or agencies bridging the gap. Debates of community versus facility-based health care are starting to shift towards building integrated health systems. But now it is shifting towards combining the strengths of both approaches in health systems.

Carl also mentioned about Community based PHC (CBPHC) and integrating CBPHC with Seed Scales. Highlighting the broad sweep of history of PHC, he underlined the amazing similarities of ancient system of India, China and Greece practicing as natural medicines, role of religions, shamans and Babylon public square concept and further added about hot and cold forms, spirits possession for ill health, humors, miasmas and

heating practices[3]. He enlightened that earlier PHC was preventive and integrated but Hippocrates differentiated medicine and public health streams. Indian systems like Charak, Susuruta, Vagbhata were based upon Ayurveda and Chopra commission legalized it but western doctors defeated it. Successful research on Indian herbs like Rauwolfia for hypertension gave the idea of earning of money to the commercial companies and similarly, yellow emperor's classic system in China.

Let us note down that it was Virchow, the german pathologist who originated modern PHC as social medicine. Later on, post world war-1, Dawson report, Peckham health centers social work centres in US strengthened it. Ding Xian from Beijing introduced the Mao's concept of Bare food doctors for a quarter of world population but later on, it was collapsed by Deng Xiao Ping's economic reforms in 1980. Second and third generation projects emerged in late 1930s to 1950s, e.g., Hydrick from Indonesia; Stamper from Croatia; major historical contribution by Rockefeller foundation, developing centers from Sri lanka, Kerala in India. In 1950s, Kark et al; Pholela-South Africa[4] developed the health system. In contrast an example of a partly successful run public health program was in South Africa. The Community Oriented Primary Care (COPC) founded by Sidney and Emily Kark in the rural area of South Africa. The organization's philosophy was comprehensive healthcare for all, which focused primarily on the health needs and using preventative care. The Kark's used a team of doctors, nurses and trained healthcare workers for community programs for mothers, children and to strictly promote infant and childhood development and growth. Another of their goal was to train community members to track diseases which were data oriented and it's goal was to keep records of births, deaths, and illnesses in the area. This model has been proven to be partly successful in South Africa due to the lack of help from the government. This lack of help was political in nature and was strongly tied to Apathied, hence resulting in the Kark's leaving South Africa. Though partly successful, this was the model that the USA emulated for their Primary Health Care guidelines. From 1960 to 1970, other health systems also came into the existence like Narangwal Punjab, Fendall from Kenya, Geiger from US and Aroles-Jamkhed and finally, in 1978 at Alma Ata. The Jamkhed model has achieved results far in excess of the Millennium Development Goals (MDG) in about 40 years. It transformed the community so much that it is not just healthier; it is also self-sufficient and economically respectable.

Let us re-assert that there are three vital pillars of PHC; Equity, intersectoral co-ordination and community participation. Empowering Community in Nagpur, Gadchiroli model in Home Based New Born Care (HBNC) Dr. Abhay Bhang resulted in lowering the Infant Mortality Rate and then Neonatal Mortality Rate to 25/1000 Live Births from 125/1000 live births. The driving force was-Think locally but act globally. The model was replicated in Pakistan, Africa and Bangladesh with equal success by the people power-so called Demographic Dividends. Certainly, achievement of high and equitable coverage of integrated primary health-care services requires consistent political and financial commitment, incremental implementation based on local epidemiology, use of data to direct priorities and assesses progress, especially at district level, and effective linkages with communities and non-health sectors. In order to demonstrate the Demographic Dividends, we are quoting another example to highlight as to how post-war Liberia was built up by Community Health Workers was vividly brought out by Dr. Panjabi. This resolved multiple problems such as providing employment, boosting the economy along with the purpose of spreading knowledge to the people.

Carl also emphasized the role of traditional practitioners and their competitions with the modern practitioners in PHC. Their roles are still ambiguous in CBPHC especially in the poorest and the remotest areas. In order to make the public health routine work interesting, he briefed out for some natural

experiment and importance of people and not their numbers. Pat Rubinstein and Massachusetts, the great old epidemiologists used to understand the shoe leather epidemiology of diseases, their causation, diagnosis, control and shared with people and work intelligently with the community. Lastly and quite importantly, he concluded the discussion with a trawling question about ways and means for qualified doctors to actually visit the villages for PHC in the developing countries.

Now we still have old health challenges and certainly, new priorities have emerged (eg, HIV/AIDS, chronic diseases, and mental health), ensuring that the tenets of Alma-Ata remain relevant. We examine 30 years of changes in global policy to identify the lessons learned that are of relevance today, particularly for accelerated scale-up of primary health-care services necessary to achieve the Millennium Development Goals, the modern iteration of the "health for all" goals. Revitalizing Alma-Ata and learning from three decades of experience is crucial to reach the ambitious goal of health for all in all countries, both rich and poor; also for today and tomorrow. Carl has rightly said," There is no universal solution, but there is a universal process to find an appropriate local solution"? [3, 5]

In 20 years from now, at the half century of Alma-Ata,we could see a different world, with basic health care reaching many of the poorest families. Wonderfully, we have a bubbling contingent of of adolescents which is one third of the total population in India, anxiously looking forward to PHC. However, to achieve this goal we need to revitalize the original revolutionary principles of Alma-Ata, sticking consistently to the core values of universal access for care, equity, community participation, intersectoral collaboration, and appropriate use of resources. We believe that revitalization of the tenets of the Alma-Ata Declaration is necessary to meet the MDGs in 2015 and beyond. Like the first primary health-care revolution, this will take champions—as Mahler said at the 2008 World Health Assembly "unless we all become partisans in renewed local and global battles for...equity...we shall indeed betray the future of our children and grandchildren."

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Effectiveness of Music Therapy on Communication Skill of Autistic Children

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Abstract

The central impairments of people with autistic spectrum disorder (ASD) include social interaction, language and communication. Music therapy uses music and its elements to enable communication and expression, thus attempting to address some of the core problems of people with ASD.

Keywords: Autism; Communication; Music therapy; Non-verbal communication; Imitation & Listening Response.

Introduction

Human communication is embodies a rich tapestry of information conveyed through elements of verbalizations, gestures and emotional expressions. It is a process by which people interact with each other to form social relationships. It serves a number of social needs such as sharing pleasure, affection, desires, feeling, attitude etc it makes our life livelier. Moreover it helps us to develop an identity to ourselves. Communication is therefore a complex ongoing process, which develops and strengthens by experiences on social interaction.

Childhood Autism, a subgroup of Pervasive developmental disorder (PDD). PDD is a well-recognized syndrome. The key construct is the "triad of impairment", which affects social interaction, language and communication, and behavior and imagination, that can be identified through examination of early development and current presentation. Autism a developmental disability, significantly affecting verbal and nonverbal communication of children.

Music therapy uses music and its elements to enable communication and expression, thus attempting to address some of the core problems of people with ASD. Music therapy has been defined as "a systematic process of intervention to help the client to promote health, using musical experiences and the relationships that develops through them as dynamic forces of change".

Music therapy also provides avenues for communication that can be helpful to those who find it difficult to express themselves in words. Research supports connections between speech and singing, rhythm and motor behavior, memory for academic material, and overall ability of preferred music to enhance mood, attention, and behavior to optimize the student's ability to learn and interact.

The processes that occur within musical improvisation may help people with ASD to develop communicative skills and their capacity for social interaction. Children with

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autism exhibit a high level of preference for the music modality, demonstrate a highly accurate memory for song lyrics, increased initiation of singing compared with speaking, and significant increases in attention, motivation, and emotional engagement during music activities.

Need for the study

Worldwide average ASD prevalence was 6.7 per 1,000 children in 2000 and 6.6 per 1,000 in 2002, or approximately 1 in 150 children. 4.5 cases per 1,000 births were reported in 2006. It means that an estimate of 67 children are diagnosed a day. The overall prevalence of autism for children between 3 and 12 years of age 1.3 cases per 1,000 births. Average male:female ratio is 4:1 and 1 in 104 males will be diagnosed.

In India, Prevalence of autism is on the rise. An estimate of 1 in every 10, 000 births were considered accurate during the 1970's, but today's literature agrees on a rate of 1 in every 150. They were 17.40 lakhs in 2005, 40 lakhs in 2006, and today the autism-affected population in India would be close to a Crore. That's 20,000 new cases every year. Unlike in the US and some European countries, there are no studies in India that document the rise in autism.

The worrisome syndrome has no treatment till now. While the disorder is not rare, a majority of children with autism, even in urban India have not been diagnosed and do not receive the services they need. In India there is a tremendous lack of awareness among professionals including medical professionals who may miss- diagnose and /or under diagnose the condition. Music therapy is emerging as one of the mode of treatment, accepted as an intervention for autistic people.

Statement of problem

A study to assess the effectiveness of music therapy on communication skill of autistic children in selected autistic center, Bangalore.

Objectives of the study

1. To assess the communication skill of autistic children among the experimental and control group in terms of pretest scores.
2. To assess the effectiveness of music therapy on communication skill of autistic children with pretest and posttest scores among experimental and control group.
3. To determine the association between pretest posttest communication skill and demographic variables among the autistic children.

Hypothesis

$H_{1:}$ There will be significant difference between the pretest and posttest communication skill scores of autistic children, among experimental group.

$H_{2:}$ There will be a significant difference between the communication skill scores of experimental and control group in terms of posttest score.

$H_{3:}$ There will be a significant association between the communication skill scores and selected demographic variables.

Conceptual framework

The conceptual framework selected for the study was based on Ernestine Wiedenbach's "prescriptive theory" (Helping art of clinical nursing theory 1968).

Methodology

The study design consisted of an evaluative approach with Quasi-experimental design. The population comprised of autistic children from selected autistic centers, Bangalore. The sample of 50 autistic children, 25 in experimental group & 25 in control group was selected by using Purposive sampling technique.

Development and description of tool

The tool developed and used for data collection was Modified observational rating scale on communication skill consisting of two sections. Section A consisted of 17 items related to demographic variables and section B consisted of 40 items related to communication skill of autistic children. Covering the following areas, Receptive language, Expressive language, Verbal response, Non verbal response, Imitation and listening response.

Scoring

The rating scales consist of 40 items, to be judged as Never, sometimes and always. The Never response was given '0' score, Sometimes response was given '1' score, and Always response was given '2' score with a total aggregate of 120 score.

Selection of music

The following steps are adopted for selection of music for the study. Development of criteria checklist, Development of music therapy schedule, Content validation of music, Pretesting of the music therapy on autistic children.

Validity and reliability of the tool

The content validity of the tool was established by 19 experts. The tool was found to be reliable and feasible. The reliability of the tool was established by using Split half method. The reliability Co-efficient is 0.9754 and Validity Co-efficient is 0.9876.

Data analysis

The data gathered were analyzed and interpreted according to the objectives. Descriptive statistics were used and Mean, median, and standard deviation with graphical presentation of data. Inferential statistics was used to test hypothesis at 0.05 level significance. Chi-square test was applied for finding relationship between communication skill and Music therapy and association between findings with demographic variables respectively.

Research findings

Section A: Description of Demographic variables (Sample characteristics)

- Majority of the respondents 25 (50%) were in the age of 10-12 years.
- Majority of the respondents 39 (78%) were male.
- Majority of the respondents 31 (62%) were not having any complication during birth.
- Majority of the respondents 25 (50%) wear begin to speak at 16-24 months of age.
- Majority of respondents 41 (82%) were from Hindu families.
- Majority of the respondents 31 (62%) were from nuclear families.
- Majority of the respondents 37 (74%) belongs to family having income of above Rs 6000 per month.
- Majority of the respondents fathers 31 (62%) and respondents mothers 36(72%) were completed degree and above.
- Majority of the respondents 39(78%) from urban area.
- Majority of the respondent's father's 20 (40%) occupation was private employee and mothers 31 (62%) were housewives.
- Majority of respondents 26 (41%) were having 4-6 persons in the home.
- Majority of respondents 21(42%) were single child.
- Majority of the respondents 39 (78%) was not having any family history of mental illness / Autism.
- Majority of respondents 24(48%) were watching television.
- Majority of respondents 17(34%) were with occupational therapy.

- Majority of the respondents 36 (72%) were not having any associated health problems.

Section B: Data on effectiveness of music therapy on communication skills of autistic children.

There is improvement in communication after music therapy among experimental group. In the aspect of Non-verbal Response pretest and post test score was 48.3% and 68.7% which is more as compare to other aspect. However, The statistical paired't' test implies that the difference in the pretest and posttest communication skill scores in various aspects found statistically significant at 5% level. Hence the research hypothesis H1 is accepted. So this indicates music therapy is effective for improvement in communication skill of autistic children.

Combined Mean Post test Score of Communication Skill among Experimental and Control Group is 79.1% and 46.5%. The obtained 't' value is 11.07. However, posttest scores of communication skill in various aspects among Experimental and Control Group found statistically significant at 5% level. Therefore research hypothesis (H_2) was accepted. So this indicates music therapy is effective in the improvement of communication skill of autistic children.

Table 1: Aspect wise Mean Pre test and Post test Communication Skill Scores of Experimental Group

n = 25

No.	Aspects	Communication Skill Scores (%)						Paired 't' Test
		Pre test		Post test		Enhancement		
		Mean	SD	Mean	SD	Mean	SD	
I	Receptive Language	48.4	10.6	80.6	8.6	32.2	8.1	19.88*
II	Expressive Language	44.4	9.4	78.6	7.7	34.2	7.6	22.50*
A	Verbal Response	43.0	8.7	79.1	7.1	36.1	8.3	21.75*
B	Non-verbal Response	48.3	9.1	68.7	10.0	20.4	10.4	9.81*
C	Imitation & Listening Response	43.8	10.2	86.1	7.3	42.3	9.9	21.36*
	Combined	45.4	8.9	79.1	7.1	33.7	8.9	18.93*

Significant at 5% level, t (0.05, 24df) = 2.064

Table 2: Comparison of Aspect wise Mean posttest scores of communication skill among Experimental Group and Control Group

No.	Aspects	Communication skill scores				Student 't' Test
		Experimental Group (n=25)		Control group (n=25)		
		Mean	SD	Mean	SD	
I	Receptive Language	80.6	8.6	53.1	12.5	9.06*
II	Expressive Language	78.6	7.7	44.3	14.1	10.68*
A	Verbal Response	79.1	7.1	42.5	13.6	11.93*
B	Non-verbal Response	68.7	10.0	47.5	14.7	5.96*
C	Imitation & Listening Response	86.1	7.3	44.6	16.3	11.62*
	Combined	79.1	7.1	46.5	12.9	11.07*

* Significant at 5% level

Table 3: Comparison of Over all Mean Posttest scores of communication skill among Experimental and Control Group

Respondents	Sample (n)	Max. Score	Communication skill scores			Student 't' Test
			Mean	Mean (%)	SD	
Experimental Group	25	120	94.96	79.1	7.1	11.07*
Control Group	25	120	55.80	46.5	12.9	

* Significant at 5% level

Table 4: Classification of Respondents on Communication Skill level of Experimental Group

Communication Skill Level	Classification of Respondents			
	Pre test		Post test	
	Number	Percent	Number	Percent
Inadequate (< 50%)	20	80.0	0	0.0
Moderate (51-75 %)	5	20.0	19	76.0
Adequate (> 75 %)	0	0.0	6	24.0
Total	25	100.0	25	100.0
Chi square	34.17*			

* Significant at 5% level, y^2 (0.05, 1df) = 3.841

Further the statistical ÷^{2} value is 34.17, which is significant at 5% level. There exists a significant association between communication skill scores of the respondents in pretest and posttest from experimental group.

Section C: Association between pretest and posttest Communication skill and Demographic Variables among Autistic Children.

Regarding gender, Male children had less communication skill scores as compared to female respondents. However there exists significant association between gender and pretest communication skill scores of autistic children. (X^2=9.03*, P<0.05).

And there exists a non-significant association between pretest communication skills scores of control group and post test communication skills scores of autistic children among experimental group & control group and other selected demographic variables such as age, religion, any birth complications, type of family, area of living, education of parents, occupation of parents, family income, language, no. of family members, No. of siblings, any psychiatric history of family, any associated health problem, behavior therapy.

Discussion

The current study findings depict a real evidence of significant difference between posttest communication skill scores of experimental group and control group. Overall mean posttest communication skill scores of Experimental and Control Group was 79.1 and 46.5%. The obtained't' value is 11.07 is statistically significant at p < 0.05 level.

The above finding was supported by C. Edgerton, study on the effect of improvisational music therapy on the communicative behaviors of autistic children. Observational study revealed significant gains in autistic children communication behaviors as measured by Checklist of Communicative Responses/Acts Score Sheet (CRASS). Commensurate decreases in scores were noted when music therapy intervention was removed.

The above finding was supported by Katherine, study on Music therapy and autism. Result shows that music therapy has proven to be a very effective method in dealing with autism, allowing individuals to build social relationships and learn how to properly behavior in social situations. Interestingly, many with autism frequently show a heightened interest in music. While they are unable to communicate verbally with others, music is an avenue for many autistic people to express themselves and communicate with others nonverbally.

Conclusion

The findings showed that none of the subjects had adequate communication skill score in the pre-test whereas 24 percent of the subjects had adequate and 76 percent of the subjects had moderate communication skill score in post-test.

From the statistical analysis it was clear that there was significant increase in the communication skill level of autistic children after administration of music therapy. From this it can concluded that music therapy was effective in bringing out communicative changes. We can use music therapy as one of the nursing intervention to treat autistic children.

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Digital Childhood: To Study the Time Spent by Children on Sunday

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Abstract

Objective: To study the time spent by children on Sunday in children aged 6 years to 12 years.

Design: Prospective study.

Setting: Paediatric department of a tertiary care teaching hospital.

Participants: Total 300 cases were included in the study. Cases were children of age group 6 years to 12 years coming to paediatric OPD during 1st June 2011 to 31st May 2012.

Methods: After informed consent, detailed history was taken and parents were asked for the details of their child's activity on Sunday.

Results: About 25% time is spent for watching television averagely, with some variation between boys & girls (boys>girls).

Conclusions: The total time spent by boys on electronic based equipments is 57.5% & by girls is 50%.

Key words: Sunday; Electronic media; TV; 6 years to 12 years.

Introduction

Recent years have seen an explosion in electronic media marketed directly at the very youngest children in our society: a booming market of videotapes and DVDs aimed at infants aged 1 to 18 months, the launching of the entire television networks specifically targeting children as young as 12 months, the development of a variety of handheld video game players for preschoolers, and a multimillion-dollar industry selling computer games for children as young as 9 months. Despite this plethora of new media aimed at the very young, little is known about children's use of such media. Thus, the total time spent by children on holiday need to be reviewed in relation to media and other factors. Few existing studies focus on the media use of children who are younger than 5. In this article, we provide a brief overview of time spent by children on Sunday in age group of 6 years to 12 years.

Methods

This prospective study was undertaken in the Department of Paediatrics during 1st June 2011 to 31st May 2012. Cases were children of age group 6 years to 12 years coming to the Paediatric OPD during the study period.

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The study included 300 children who were aged 6 years to 12 years. Parents of these children were asked about how their children spent their Sunday. Of those 300 parents, 284 parents responded well in the subsequent visit and provided the details of the activities which their children do on Sunday. Therefore, the sample used comprised information for 284 children.

Parents were asked to report what their child did the following on the Sunday: watched television, watched a video or DVD, played video games on a console, played hand-held video games or mobile games, played computer games, used the computer for something other than games like internet, read story books or news papers, did painting, listened to music, went for tuitions, did homework, slept, chatted with their parents or grandparents, played indoor or outdoor games, went for friend's birthday. Parents were then asked to report on the amount of time their children spent using these various activities on the Sunday.

Also, parents were asked about the details of their family like total number of television sets in their home, availability of television set in bedroom or child's room. Data were entered in MS Excel, cleaned and completeness checked.

Results

Total 300 cases coming to the Paediatrics OPD between age group 6 years to 12 years were studied. Out of those 300 cases, 284 gave proper feedback. In those 284, 154 were boys & 130 were girls. 8 hours of night sleep is considered for both boys & girls. Rest 16 hours were studied for the activities & the average distribution of their day-time showed as per given in table no. 1. In those 16 hours, averagely 1 hour is spent on day-to-day activities like bathing, tea, lunch, dinner, etc. About 25% time is spent for watching television averagely, with some variation between boys & girls (boys>girls). The total time spent by boys on electronic based equipments is 9 hours 12 minutes (57.5%) & by girls is 8 hours (50%).

Table 1

Activity	Average Time Spent (hrs:min)	
	Boys	Girls
Watching television	04:30	03:44
Watching a video/ DVD/ movie	00:40	00:23
Playing video games on a console	01:04	00:39
Playing hand-held video games or mobile games	00:43	00:38
Playing computer games	00:54	00:50
Using computer for something other than games like internet	00:35	00:31
Reading story books/news papers	00:30	00:44
Painting	00:18	00:50
Listening to music	00:46	01:15
Tuitions	00:45	00:50
Doing homework	01:05	01:15
Sleeping	00:45	00:50
Chatting with their parents or grandparents	00:31	00:35
Playing indoor games (chess, carom, etc.)	00:44	00:50
Playing outdoor games	00:50	00:30
Going for friend's birthday	00:20	00:30
Day-to-day activity (bath, tea, lunch dinner, etc.)	01:00	01:00
Total	**16:00**	**16:00**

Table 1

Parameter	Yes	No
Number of televisions: =2	42	242
Bedroom television	28	256
Access to either cable- or satellite-based channels	225	57
Video game console	140	144
Hand-held video game or mobile games	227	256
Computer	85	146
Internet access	28	200

The other parameter study shows that most of them are living in homes with at least 1 of each product; nearly 15% live in home with e"2 televisions. 80% children in this age live in homes with access to either cable- or satellite-based channels. Approximately 50% of the households had a video game console, and about 80% had access to a hand-held video game or mobile games. Even more households (30%) reported having a computer, and 10% of all households have internet access. Although none of this is perhaps surprising, we did find it surprising that many of these young children have televisions in their own bedrooms. This was true for almost 10% of children. The most common reason that parents named for having a television in their child's bedroom was that it frees up other television in the house so that other family members can watch their own.

Discussion

The impetus for this study came from the striking lack of empirically based knowledge about the extent of exposure to and use of media and technology by young children. This study provides the most recent information available regarding the extent of media use and media access of school going children between age group 6 years to 12 years.

In addition, the results of this survey make it clear that young children today are growing up in a media-saturated environment. For this cohort of children and presumably for future cohorts as well, access to and use of media have become part of the fabric of their daily lives.

Much has been made of the vast array of media and technology that are used by children today, and it is clear that they have far more choices available to them than previous generations. However, it is also true that this study indicates that children's use of electronic media is still, by and large, dominated by television. Also, young children do play video games or use the computer. The use of these media is for as same duration as they spend for watching television, which is not a trivial amount of time in a young child's day.

To state that television and DVD ownership was virtually ubiquitous in the homes of young children does not adequately capture the extent of media saturation in these homes. Overall, it seems clear that these children will be very different from previous generations of children with respect to their comfort with technology and the extent to which they use all forms of technology in their daily lives. An environment that directly exposes children to television—that is, being in a constant television household and having television in the child's bedroom—is related to higher chances of decreasing outdoor games.

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Evolution of Child Birth

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Abstract

Before the 1920's, birth took place at home and was attended by doctors or midwives. In the 20's, women flocked to hospitals for the 'new' modern methods of 'painless' childbirth by sedating mother. Not all doctors saw comatose childbirth as a positive step for Mom and Baby. Dr. Grantley Dick-Read of England saw the beauty in participatory childbirth. He noticed that women who had someone with them to explain events had significantly less pain. In the 1930's he wrote "Birth Without Fear". In the late 40's, the Maternity Center of New York sponsored a grant to study the effect of his methods and to allow babies to 'room in'. Fernande Lamaze, a French obstetrician, studied Russian techniques of conditioned response to reduce childbirth pain. The organization of the LaLeche League gave us the movement to promote breastfeeding.After that comes the concept of PREPARED CHILD BIRTH which gives Mom choices as to how to have her baby and along with her partner allows her to make these choices based on information not fear or ignorance. In the 60's, Dr. Robert Bradley introduced the radical concept of fathers in the delivery room. Michael Odent brought the concept of water birth.

Key words: Labor; Home deliveries; Hospital deliveries; Water birth.

Introduction

In the developing world, even today, perhaps delivery is the commonest event where life and death stand side by side for both, the expectant mother and her forthcoming newborn. India alone accounts for nearly 20% of the global burden of both maternal and child deaths against about 16% of its share in world population[1]. Institutional deliveries range from 11% to 95% in different states of India with an average of 41%[2].

There are no descriptions of childbirth in the Bible. But we know what happened in surrounding countries and that can be a guide. In the pre antibiotic era they used brightly painted birthing bricks to stand or kneel on over a scooped out hole, or they sat on a birth-stool/chair.

The woman giving birth was surrounded by women she knew and trusted – her relatives and friends. Major advances in asepsis & labor began with the introduction of hand-washing by Semmelweis in 1847 at the Vienna Maternity Hospital. Ignaz Philipp Semmelweis (July 1, 1818 – August 13, 1865) (born Ignác Fülöp Semmelweis)[21] was a Hungarian physician now known as an early pioneer of antiseptic procedures & described as the "savior of mothers"[22] . The First Clinic at the Vienna Maternity Hospital were attended by medical students who moved straight from the necropsy room to the delivery

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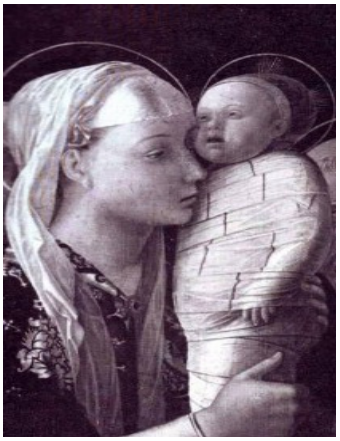
Woman giving birth on a birthing chair .

Baby was wrapped in coarsely woven linen strips, swaddling bands to promote strong, straight bones as the baby grew.



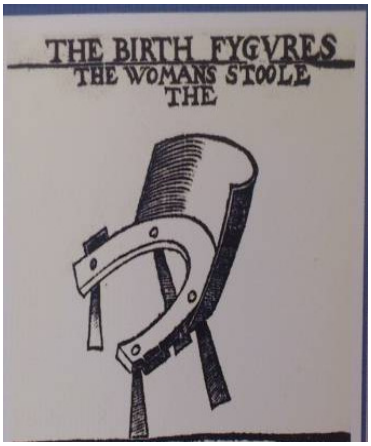
An Egyptian birthing brick; the original painted image shows the goddess Hathor presenting a child to its mother

suite. The Second Clinic were attended by midwives and midwifery students who had no contact with the necropsy room. The incidence of maternal death was as high as 18% in the first department, with puerperal fever the main cause, but only 2% in the second. Semmelweis observed that a colleague, Jakob Kolletschka, died from an illness similar to puerperal fever after being accidentally cut during a necropsy. He concluded that the infecting particles responsible for puerperal fever came from cadavers and were transmitted by hand to women attended by medical students in the first department. He therefore instituted hand disinfection with chlorinated lime (modern calcium hypochlorite, the compound used in today's common household chlorine bleach solution) for those leaving the necropsy room, after which maternal morbidity in the first department fell to the levels achieved by the second department. Later Lister's introduction of antisepsis with carbolic spray in the 1870s, based on the germ theory of Pasteur, was an important step forward in the

Model of pelvis used in the beginning of the 19th century to teach technical procedures for a successful childbirth [4]

prevention and limitation of infection a major cause of maternal mortality[23].

Birthing chair by Thomas Raynalde



Sculpture of goddess Lajja Gouri from caves of North Karnataka

Ancient Indian art Sculpture from Indian cave

Birth positions

Standing or walking, Rocking, Squatting, Birthing balls, Sitting, Kneeling over a chair, Hands and knees, Side-lying .

We find attitudes and beliefs about obstetrics more familiar to the modern world.(19). Soranus (physician) who made a significant contribution to labor begins his discussion of childbirth with a description of the good midwife.

Midwifes

Their duties were not as clear-cut as those of a modern midwife. They were expected to

- provide comfort, pain relief and encouragement to the woman giving birth
- perform rituals and prayers that would protect the woman and her baby, and keep harmful forces away
- use her expertise to birth the baby and deliver the afterbirth
- deal with problems or complications during the birth

supervise aftercare for mother and baby

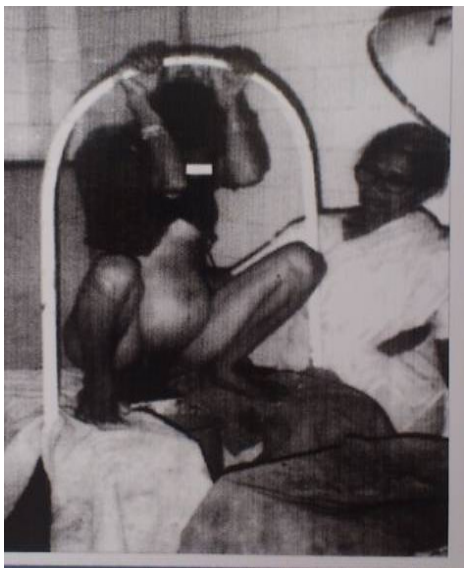
Soranus's references to other medical writings also indicate that obstetrical practice was not limited to midwives; a male physician might attend particularly difficult births. The equipment used was midwife's chair. In the seat of the chair was a crescent-shaped hole through which the baby would be delivered. Midwife used to ease the labor pains with gentle massage, with a cloth soaked in warm olive oil laid over the abdomen and genital area, and with the equivalent of hot-water bottles- bladders filled with warm oil- placed against the woman's sides. For the actual delivery, the midwife needs three assistants to stand on both sides of the chair and at the back.There was no concept of episiotomy.

At the onset of the Industrial Revolution in the 19th century, giving birth at home became more difficult due to congested living spaces and dirty living conditions. This drove urban and lower class women to newly available hospitals, while wealthy and middle-class

women continued to labor at home[9].The ability to labor without pain was part of the early feminist movement[10]. With this change from primarily homebirth to primarily hospital birth came changes in the care women received during labor: although no longer the case, in the 1940s it was common for women to be routinely sedated and for babies to be delivered from their unconscious mothers with forceps with episiotomy (termed by Dr. Robert A. Bradley as "knock-em-out, drag-em-out obstetrics"). Along with shaving of the mother's pubic region; mandatory intravenous drips; enemas; hand strapping of the laboring women; and the 12 hour monitoring of newborns in a nursery away from the mother.

Ancient Indian birthing position along with midwife .

Traditional vertical squatting parturition postur e on birth chair cum table .



Hospital birth

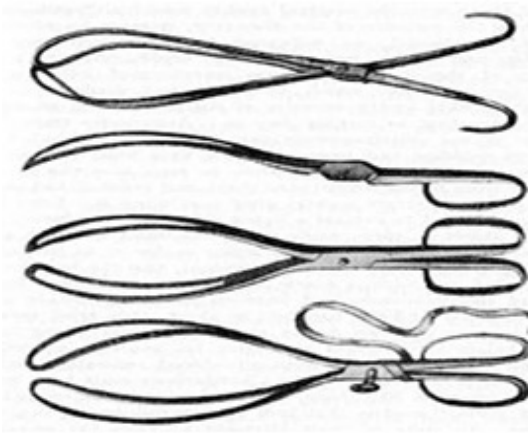
Advantages: safest childbirth environment for high risk pregnancies, emergency cesarean section facility, pediatricians and newborn medical technology is available,as also anesthesiologists provide pain relief, from epidurals to narcotics.

Rate of instrumental deliveries like forceps, ventouse increased. The introduction of obstetric forceps was of great interest to man-midwives who were distressed by the loss of live fetuses in obstructed labours. The obstetric forceps, allowing during birth, the extraction of a *living* child, was invented by the eldest son of the Chamberlen family of surgeons named Pierre (Peter). Four generations of the Chamberlen family are credited with the development of obstetric forceps, which they maintained as a secret until 1730.11(Q). In fact, the instrument was kept secret for 150 years by the Chamberlen family, although there is evidence for its presence as far back as 1634.The Chamberlen family's forceps were based on the idea of separating the two branches of "sugar clamp". In the interest of secrecy, the forceps were carried into the birthing room in a lined box and would only be used once everyone was out of the room and the mother blindfolded[20]. Hughes Chamberlen, Grand nephew of Peter the eldest, tried to sell the instrument in Paris in 1670, but the demonstration he did in front of François Mauriceau, responsible for Paris Hotel-Dieumaternity, was a resounding failure which resulted in the death of mother and child. The secret may have been sold by Hughes Chamberlen to Dutch obstetricians at the start of the 18th century in Amsterdam. Earliest mention of instrumental delivery in

Vedic era - "Ankush". Albucasis described forceps with teeth on the inner surface for dead foetus. The Chamberlain family used four pairs of forceps of different sizes with only cephalic curve.

- **Levret (1747)-**introduced the pelvic curve
- **Smellie (1751)-**reinforced pelvic curve & introduced English lock and used in aftercoming head.

Chamberlen forceps (Maldon)



- **Tarnier (1877)-**introduced axis traction.
- **Barton and Kjielland -** introduced the two specialized forceps.

Thereafter, the knowledge of forceps became widespread, with the development of models of the short straight forceps and the long forceps with a pelvic curve.The history of obstetrical forceps is long and, often, colorful. Sanskrit writings from approximately 1500 BC contain evidence of single and paired instruments; Egyptian, Greek, Roman, and Persian writings and pictures refer to forceps that were originally used for extraction following fetal demise to save the mother's life. In the last decades, however, with the ability to perform a cesarean section relatively safely, and the introduction of the ventouse or vacuum extractor, the use of forceps and training in the technique of its use has sharply declined.

When the anesthetic effects of ether and chloroform were discovered in the mid 1800's, many members of the British clergy argued that this human intervention in the miracle of birth was sin against the will of God. . According to Scripture, childbirth pain originated when God punished Eve and her descendants for Eve's disobedience in the Garden of Eden. The first use of modern anesthetic for childbirth occurred a scant 3 months after Morton's historic demonstration of the anesthetic properties of ether at the Massachusetts General Hospital in Boston, Massachusetts. James Young Simpson used diethyl ether to anesthetize a woman with a deformed pelvis for childbirth. Queen Victoria undaunted by the clergy chose one day to use an anesthetic during labor and the clergy's position crumpled like the great wall of 'Berlin'. The first woman anesthetized for childbirth in the United States was Fanny Longfellow, wife of the American poet Henry Wadsworth Longfellow. Anesthetics were subsequently used increasingly for labor pain, and the concurrent drop in mortality and morbidity in both mother and infant were attributed. In 1941, Robert Andrew Hingson (1913–1996) and Waldo B. Edwards developed the technique of continuous caudal anaesthesia using an indwelling needle[18]. The first use of continuous caudal anaesthesia in a labouring woman was in 1942[19].

Beginning in the 1940s, childbirth professionals and mothers began to challenge the conventional assumptions about the safety of medicalized births. Physicians Michel Odent and Frederick Leboyer and midwives such as Ina May Gaskin promoted birthing centers,water birth, and homebirth as alternatives to the hospital model.

Birthing centre

Presents a simulated home-like environment. Located on hospital grounds or "free standing" (i.e., not hospital-affiliated). Accept women with low-risk pregnancies — which mean that you'll have to be evaluated for risk first. No epidurals. During a medical

emergency, you may have to be transported to a nearby hospital.

The Bradley Method of natural childbirth (also known as "husband-coached childbirth") is a method of natural childbirth developed in 1947 by Robert A. Bradley, M.D. (1917–98) and popularized by his book Husband-Coached Childbirth, first published in 1965. The Bradley Method teaches couples to manage labor through deep breathing and the support of a partner or labor coach[5]. It relies heavily on training fathers to be labor "coaches," or partners.

Water birth

Baby spends nine months doing water ballet in a warm pool of amniotic fluid, and then makes a sudden, harsh entrance into the cold, bright (and dry) world. Michael Odent, another French physician, put mother and baby both in the water. This appears to help some women ease labor pain. This reduces a baby's stress. If you're opting for a water birth, you'll labor in a warm tub or pool, kept at approximate body temperature (95 to about 100 degrees, but no more than 101, because your body temperature could rise, causing the baby's heart rate to increase). Since a baby's breathing will not start until he or she comes out of the water and into the air (babies don't breathe in utero), drowning is not considered a risk of water births. For a couple of reasons, however, a baby's underwater entry should be limited to no more than a few moments (ten seconds is the norm in the U.S.): first, because the umbilical cord can tear, cutting off the baby's oxygen lifeline, and second, because once the placenta separates from the uterus — which can happen at any time after delivery — it can no longer provide the baby with sufficient oxygen.A special underwater Doppler device will monitor your baby's heartbeat. You can also receive medications through an IV while you are in the water.

Coffin birth

Known in academia by the more accurate term postmortem fetal extrusion [11,12] is the expulsion of a nonviable fetus through the vaginal opening of the decomposing body of a pregnant woman as a result of the increasing pressure of intra abdominal gases. Typically, as a dead body decomposes, anaerobic bacteria in the gastrointestinal tract proliferate and release gases such as carbon dioxide, methane, and hydrogen sulfide [13,14] . As the volume of gas increases, the pressure begins to force various body fluids to exude from all natural orifices[9]. It is at this point during the decomposition of a pregnant body that amniotic membranes become stretched and separated, and intra abdominal gas pressure may force the eversion and prolapse of the uterus, which would result in the expulsion of the fetus through the vaginal canal[15]. The earliest presented case occurred in the year 1551 when a pregnant woman was tried and hanged by the courts of the Spanish Inquisition. Four hours after her death, and while the body still hung by the neck, two dead infants were seen to fall free of the body[16].

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Patau Syndrome

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Abstract

Patau syndrome is a chromosomal abnormility in which patient has an additionalchromosome 13 due to non disjunction of chromosome during meiosis. Some are caused by translocation while others are caused by mosaic patau syndrome.

Key words: Patau syndrome; Trisomy 13; Trisomy D.

Introduction

Patau syndrome is a chromosomal abnormility in which patient has an additional chromosome 13 due to non disjunction of chromosome during meiosis. A small percentage of cases occur when only some of the body's cells have an extra copy of chromosome 13, resulting in a mixed population of cells with differing numbers of chromosomes. This is called Mosaic Patau.

Case summary

A 22 years old rural habitat $G_2P_1L_1A_0$ female was admitted to our labor room with complaints of pain in the lower abdomen. The pain was continuous, dull aching, progressive, radiating to lower back and medial aspect of thigh. She had history of fever 3 days back, for which she took treatment details of which are not available. She was referred herein by a private practitioner.

According to her last menstrual period, her gestational age was 29 weeks. She was a registered case at PHC with 3 antenatal visits. No antenatal USG was done at PHC. She had taken iron and folic acid tablets with 2 doses of TT injection. She had family history of third degree consanguineous marriage. Her first child is a three year girl who was a full-term normal vaginal delivery without any congenital anomaly.

On examination, she was an averagely built lady; with pulse 80/min & blood pressure-120/70 mmHg. Mild pallor was present. Head circumference was 23 cm (<3rd percentile). Per abdominal examination showed uterus of 26 weeks and a fetus with cephalic presentation. Per vaginal examination showed 1 finger loose dilated cervix without any PV bleeding. USG showed live fetus of 26 weeks and 2 days gestational age with congenital anomaly with holoprosencephaly variant with hypotelorism and moderate polyhydroamnios.

Mother's investigation revealed Hb 10 gm%, TLC-11000, Platelet Count-2 lakhs/Cmm. Liver function tests and renal function tests

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Table 1

Clinical Features	Patau Syndrome	Edward Syndrome	Our Patient
Close set eyes	+	-	+
Microcephaly	+	+	+
Holoprocencephaly	+	-	+
Low set ears	+	+	+
Small lower jaw	+	-	+
Hypotelorism	+	-	+
Polydactyly	-	+	-
Inguinal hernia	-	+	-
Omphalocoele	+	+	-
Congenital heart defects	+	+	Can't be Evaluated
Underdeveloped Brain	+	+	+
Cleft lip/palate	+	+	-
Rocker Bottom Feet	-	+	-
Clenched hand with overlapping of fingers	+	+	+
Single umbilical artery	+	+	-

were within normal limits. VDRL, HbsAg and Tridot were negative. Urine showed traces of albumin. She delivered IUD fetus by preterm vaginal delivery.

The baby was an IUD fetus, with multiple congenital anomalies, listed in table no.1.

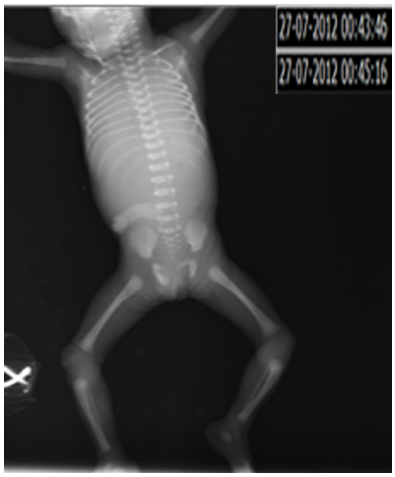
From above table we can rule out Edward syndrome. Ophthalmology opinion was taken which states presence of 2 orbits.

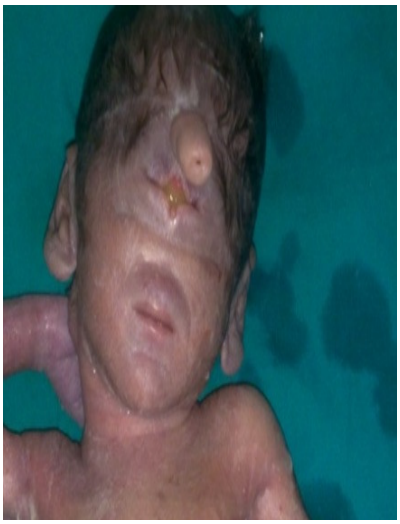
Following investigations were done.

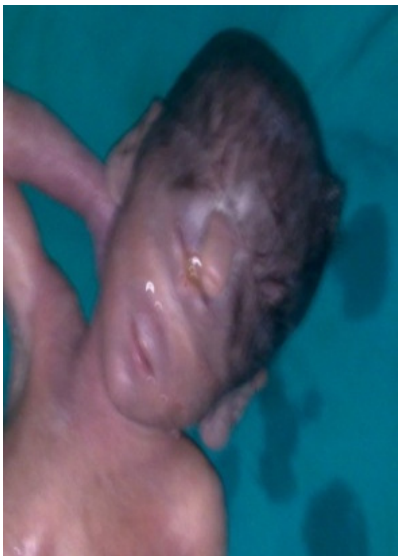
USG Abdomen: Malrotated organs; B/L polycystic kidney.

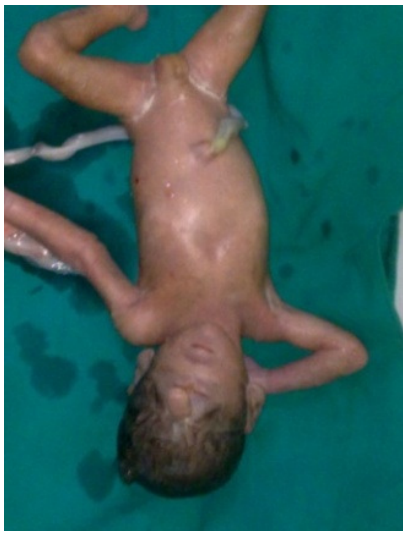
USG Cranium: Underdeveloped Brain

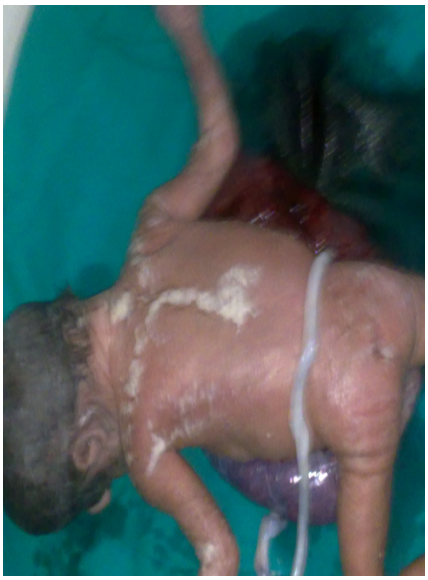
CT Scan Brain (plain): Microcephaly with absent septum pellucidum & a single large ventricle in brain surrounded by thin rim of cortex showing pachygyria. The thalami were partially separated by a rudimentary third ventricle. The small partly formed interhemispheric fissure and falx were seen only in high parietal region & were absent anteriorly Findings were suggestive of holoprosencephaly.

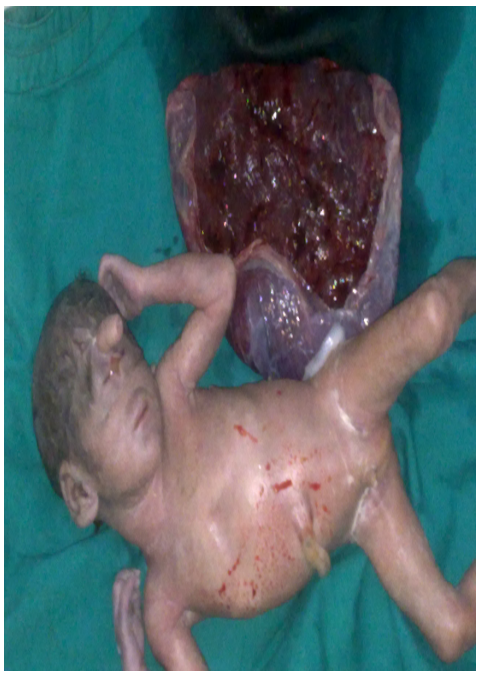












Discussion

Patau syndrome, or "Trisomy 13", as it was first called, was first observed by Thomas Bartholin in 1657. However, the actual genetic and chromosomal-related parts of it were discovered by Dr. Klaus Patau in 1960, hence the name "Patau syndrome". It affects about 1 in 12,000 live births. More than 80% of infants with Patau syndrome die within their first year of life.

Patau syndrome, also known as trisomy 13 and trisomy D, is a chromosomal abnormality, a syndrome in which a patient has an additional chromosome 13 due to a nondisjunction of chromosomes during meiosis. Some are caused by Robertsonian translocations, while others are caused by mosaic Patau syndrome. The extra chromosome 13 disrupts the normal course of development, causing heart and kidney defects. Like all nondisjunction conditions (such as Down syndrome and Edwards syndrome), the risk of this syndrome in the offspring increases with maternal age at pregnancy, with about 31 years being the average.

Dr. Klaus Patau

Genetics

Patau syndrome is most often the result of trisomy 13, meaning each cell in the body has three copies of chromosome 13 instead of the usual two. Patau syndrome can also occur when part of chromosome 13 becomes attached to another chromosome (translocated) before or at conception in a Robertsonian translocation. Affected people have two copies of chromosome 13, plus extra material from chromosome 13 attached to another chromosome. With a translocation, the person has a partial trisomy for chromosome 13 and often the physical signs of the syndrome differ from the typical Patau syndrome.

Most cases of Patau syndrome are not inherited, but occur as random events during the formation of reproductive cells (eggs and sperm). An error in cell division called non-disjunction can result in reproductive cells with an abnormal number of chromosomes. For example, an egg or sperm cell may gain an extra copy of the chromosome. If one of these atypical reproductive cells contributes to the genetic makeup of a child, the child will have an extra chromosome 13 in each of the body's cells. Mosaic Patau syndrome is also not inherited. It occurs as a random error during cell division early in fetal development.

Patau syndrome due to a translocation can be inherited. An unaffected person can carry a rearrangement of genetic material between chromosome 13 and another chromosome. This rearrangement is called a balanced translocation because there is no extra material from chromosome 13. Although they do not have signs of Patau syndrome, people who carry this type of balanced translocation are at an increased risk of having children with the condition.

Manifestation

Of those fetuses that do survive to gestation and subsequent birth, common abnormalities include:

Nervous system

Mental and motor challenged, Microcephaly, Holoprosencephaly (failure of the forebrain to divide properly).

Eye Defects

Microphthalmia, Peters anomaly (a type of eye abnormality), cataract, iris and/or fundus (coloboma), retinal dysplasia or retinal detachment, sensory nystagmus, cortical visual loss, and optic nerve hypoplasia.

Spinal defect

Meningomyelocele.

Musculoskeletal and cutaneous defects

Polydactyly (extra digits), Low-set ears, Prominent heel, Deformed feet known as rocker-bottom feet, Omphalocele (abdominal defect), Abnormal palm pattern, Overlapping of fingers over thumb, Cutis aplasia (missing portion of the skin/hair), Cleft palate.

Urogenital defects

Abnormal genitalia, Kidney defects (polycystic kidney).

Other defects

Heart defects (ventricular septal defect), Single umbilical artery.

Recurrence risk

Unless one of the parents is a carrier of a translocation the chances of a couple having another trisomy 13 affected child is less than 1% (less than that of Down syndrome).

Treatment

Medical management of children with Trisomy 13 is planned on a case-by-case basis and depends on the individual circumstances of the patient. Treatment of Patau syndrome focuses on the particular physical problems with which each child is born. Many infants have difficulty surviving the first few days or weeks due to severe neurological problems or complex heart defects. Surgery may be necessary to repair heart defects or cleft lip and cleft palate. Physical, occupational, and speech therapy will help individuals with Patau syndrome reach their full developmental potential.

Prognosis

More than 80% of children with Patau syndrome die within the first year of life.

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Marfan's Syndrome

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Abstract

Marfan's syndrome is a genetic disorder of the connective tissue. People with Marfan's tend to be unusually tall, with long limbs and long, thin fingers. The syndrome is inherited as a autosomal dominant trait, carried by the gene FBN1, which encodes the connective protein fibrillin-1.

Key words: Marfan's syndrome; Connective tissue disorder; FBN1; Fibrillin-1.

Introduction

Marfan's syndrome is an inherited connective tissue disorder that is transmitted as an autosomal dominant trait. People have a pair of FBN1 genes. Because it is dominant, people who have inherited one affected FBN1 gene from either parent will have Marfan's syndrome. Hence, one affected parent is sufficient to pass on the disorder to the child. Being a connective tissue disorder, Marfan's syndrome affects almost all of the body's systems, including the skeletal, cardiovascular, nervous, skin, and pulmonary systems.

Case summary

Eleven years boy brought by his parents to our OPD with complaints of diminishing of vision since birth. There was no history of trauma to eyes. Also he was not a known case of juvenile diabetes mellitus or juvenile hypertension. There was no history of any other eye complaint, delayed milestones, any surgery, respiratory illness, cardiac illness. In family history, his father was also suffering from similar complaints along with some cardiac problem and long limb deformities, details of which were not available. His father died 6 years ago in same course of illness. Also his paternal grandmother was suffering from similar illness & she expired in same course. His parents had family history of non-consanguineous marriage. He was a full term normal hospital delivery with good cry and adequate weight at birth. Our case was first issue of parents out of their 3 siblings (2 males & a female); none of them having any congenital anomaly or similar illness. We admitted the boy to our hospital in paediatric ward for further investigations & management.

On examination, his vital parameters were normal as per his age. In anthropometric examination, he was having extra long upper limbs & lower limbs. His head circumference was 52 cm, chest circumference – 59 cm, height – 140cm, upper segment /lower segment ratio – 0.70 (reduced), arm span –

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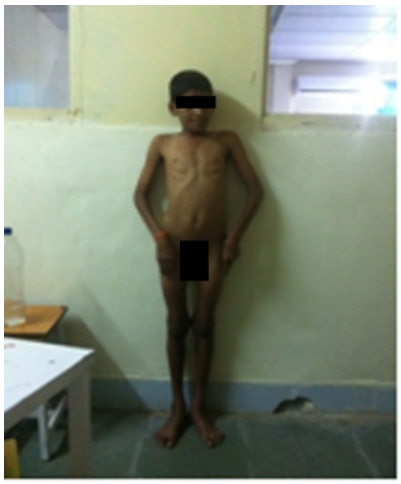
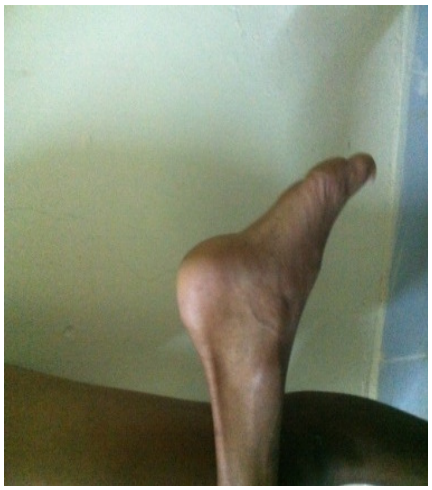
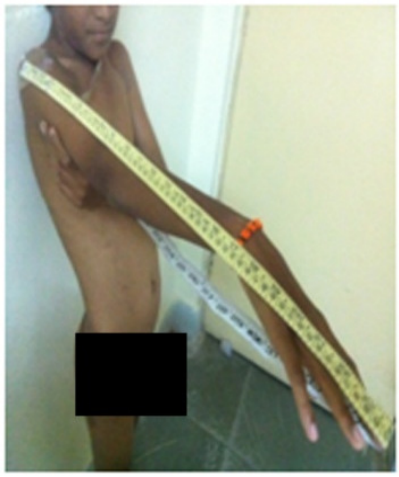
Reduced elbow extension



Hind-foot deformity

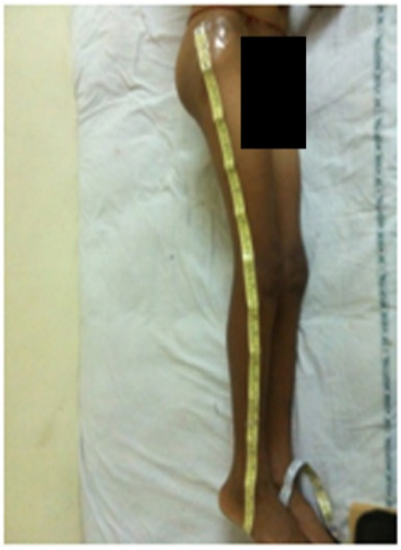


Long upper limb



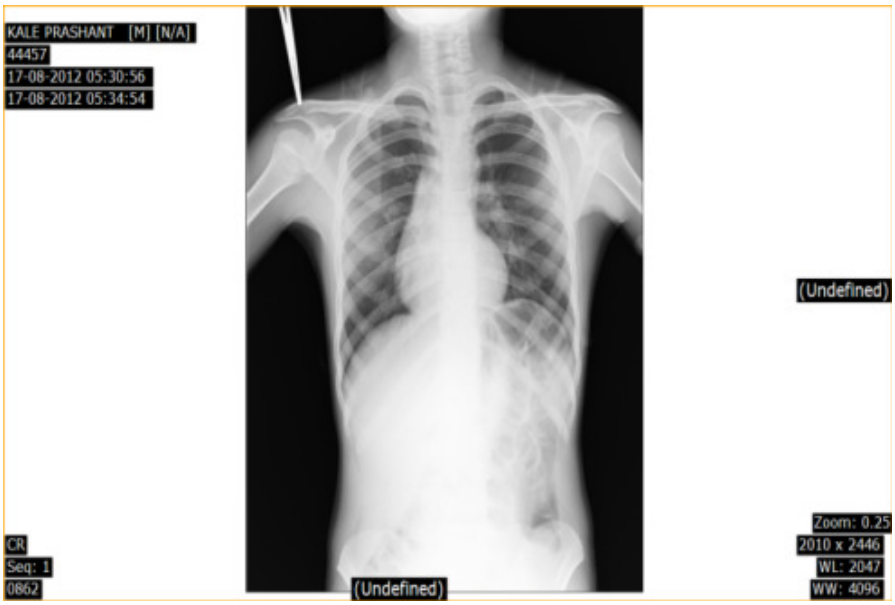
Thumb sign

Long lower limb

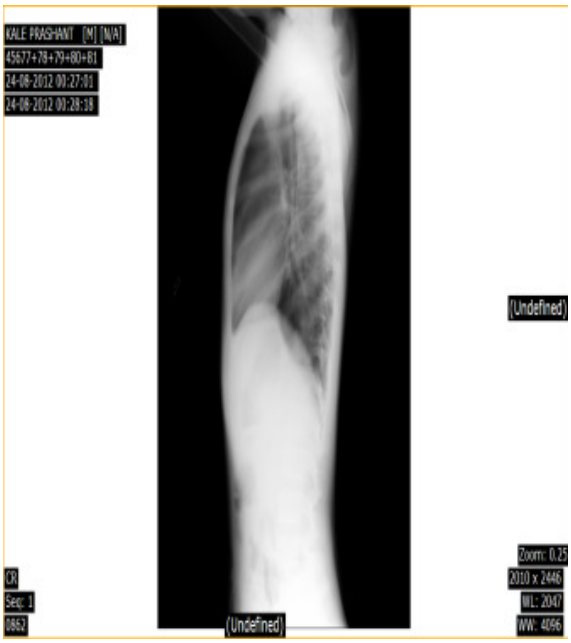


Wrist sign

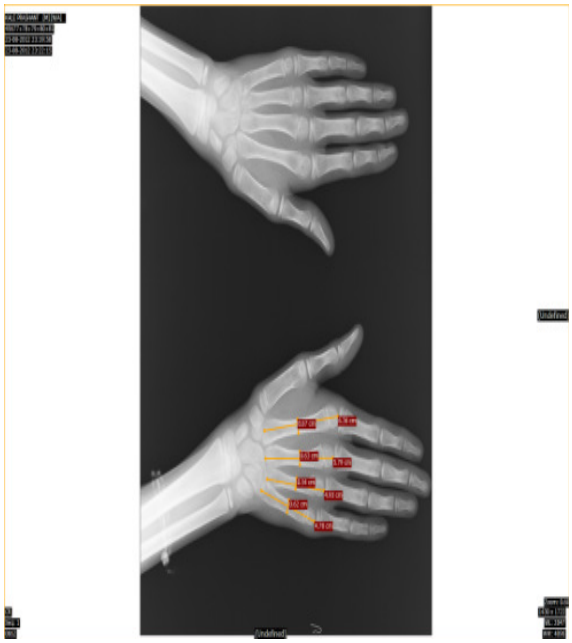
Chest x-ray PA view



Chest x-ray lateral view



X-ray showing long metacarpals

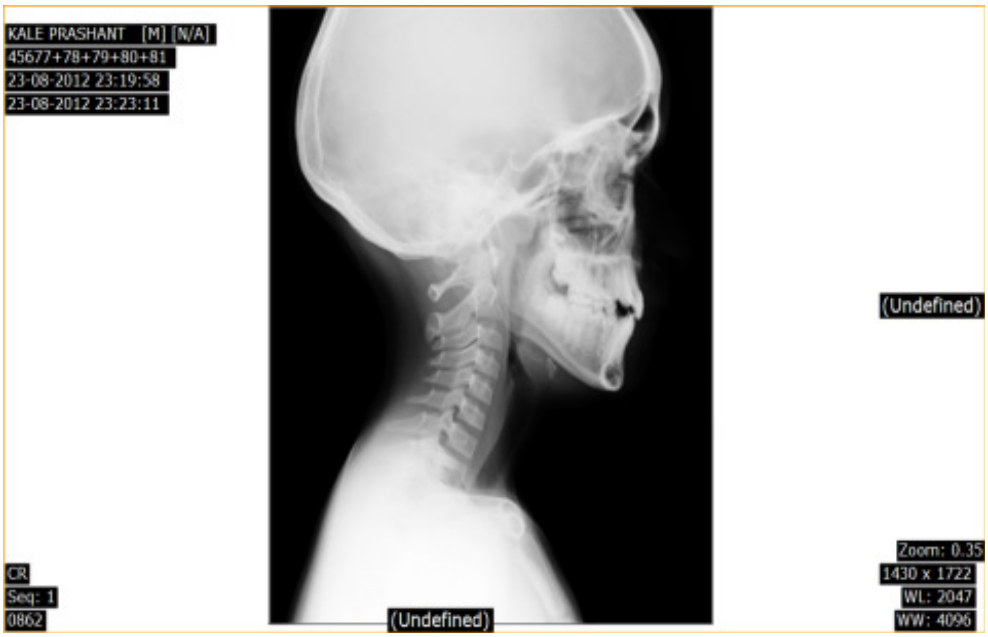


142cm, mid arm circumference – 16 cm & weight – 25.03 kg. All systemic examinations were within normal limits. Opinion from Ophthalmologist was taken which after fundoscopy stated that his vision was 6/36 for both the eyes & there was superotemporal displacement of lenses in both the eyes. No retinal detachment was there. There was no any spine deformity like scoliosis or chest deformity present.

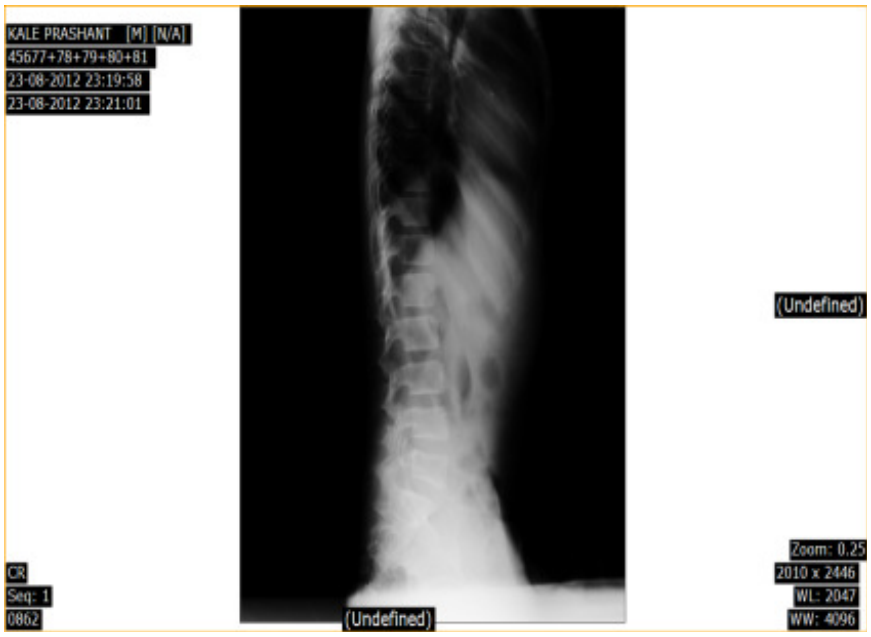
The following investigations were done:

Hb – 10.5 gm%; TLC – 6900/cmm; (N– 46, L – 42, E – 07, M – 05.)

X-ray skull



X-ray Spine



Serum electrolytes:

Sodium – 138 mmol/L. Potassium – 4.4 mmol/L. Calcium – 9.8 mg%.

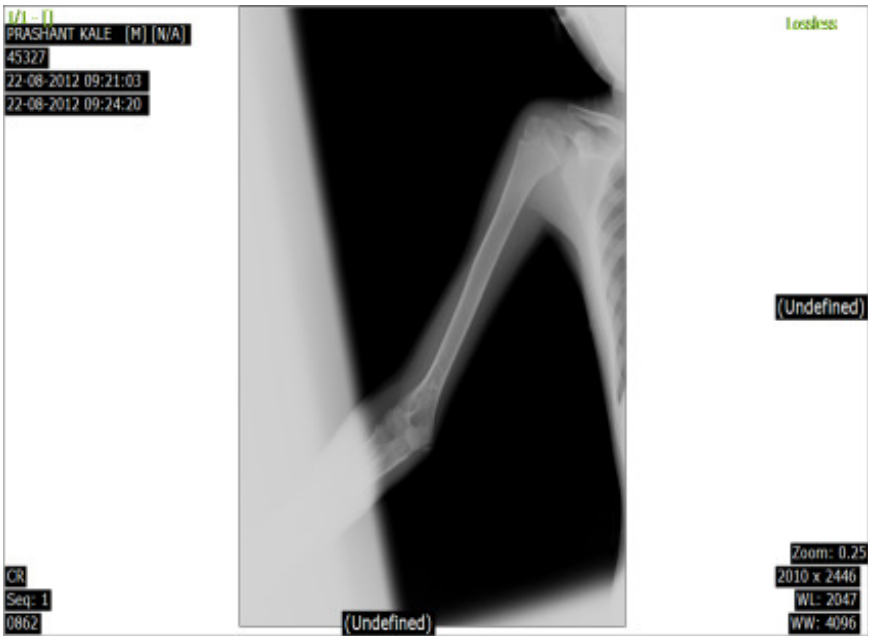
Urine examination was normal.

X-ray chest was normal.

2D ECHO didn't show any cardiac anomaly.

Colour Doppler of aorta didn't reveal any abnormality.

X-ray long bone (right arm)



We diagnosed this as Marfan's syndrome on following basis:

- Family history of Marfan's syndrome was present.
- Superotemporal displacement of lens.
- Systemic Score: Total = 8.
  - Wrist and thumb sign = 3
  - Hind-foot deformity = 2
  - Reduced elbow extension = 1
  - Myopia >3 diopters = 1
  - Reduced upper segment/lower segment ratio & increased arm/height & no severe scoliosis = 1

Discussion

Marfan's syndrome is a genetic disorder of the connective tissue with autosomal dominant inheritance. People with Marfan's tend to be unusually tall, with long limbs and long, thin fingers. Marfan's syndrome is named after Antoine Marfan, the French paediatrician who first described the condition in 1896. The gene linked to the disease was first identified by Francesco Ramirez in 1991.

Marfan's syndrome has a range of expressions, from mild to severe. The most serious complications are defects of the heart valves and aorta. It may also affect the lungs, the eyes, the dural sac surrounding the spinal cord, the skeleton and the hard palate.

Marfan's syndrome affects males and females equally, and the mutation shows no ethnic or geographical bias. Estimates indicate about one in 3,000 to 5,000 individuals have Marfan's syndrome.

Signs and Symptoms

The constellation of long limbs, dislocated lenses and the aortic root dilation are generally sufficient to make the diagnosis of Marfan's syndrome with reasonable confidence.

Skeletal System

Most of the readily visible signs are associated with the skeletal system. Many individuals with Marfan's syndrome grow to above-average height. Some have long, slender limbs (dolichostenomelia) with long fingers and toes (arachnodactyly). An individual's arms may be disproportionately long, with thin, weak wrists. In addition to affecting height and limb proportions, Marfan's

syndrome can produce other skeletal anomalies. Abnormal curvature of the spine (scoliosis), abnormal indentation (pectus excavatum) or protrusion (pectus carinatum) of the sternum are not uncommon. Other signs include abnormal joint flexibility, a high palate, malocclusions, flat feet, hammer toes, stooped shoulders, and unexplained stretch marks on the skin. It can also cause pain in the joints, bones and muscles in some patients.

Eyes

Marfan's syndrome can also seriously affect the eyes and vision. Nearsightedness and astigmatism are common, but farsightedness can also result. Subluxation (dislocation) of the crystalline lens in one or both eyes (ectopia lentis) also occurs in 80% of patients. In Marfan's syndrome, the dislocation is typically superotemporal whereas in the similar condition homocystinuria, the dislocation is inferonasal. Sometimes eye problems appear only after the weakening of connective tissue has caused detachment of the retina. Early

Lens dislocation in Marfan's syndrome;

The lens was kidney-shaped and was resting against the ciliary body

onset glaucoma can be another related problem.

Cardiovascular System

The most serious signs and symptoms associated with Marfan's syndrome involve the cardiovascular system: undue fatigue, shortness of breath, heart palpitations, racing heartbeats, or angina pectoris. Cold arms, hands and feet can also be linked to Marfan's syndrome because of inadequate circulation. A heart murmur, abnormal reading on an ECG, or symptoms of angina can indicate further investigation. The signs of regurgitation from prolapse of the mitral or aortic valves result from cystic medial degeneration of the valves, which is commonly associated with Marfan's syndrome. However, the major sign that would lead a doctor to consider an underlying condition is a dilated aorta or an aortic aneurysm. Sometimes, no heart problems are apparent until the weakening of the connective tissue (cystic medial degeneration) in the ascending aorta causes an aortic aneurysm or aortic dissection, a surgical emergency. An aortic dissection is most often fatal and presents with pain radiating down the back, giving a tearing sensation.

Lungs

Marfan's syndrome is a risk factor for spontaneous pneumothorax. In spontaneous unilateral pneumothorax, air escapes from a lung and occupies the pleural space between the chest wall and a lung. The lung becomes partially compressed or collapsed. This can cause pain, shortness of breath, cyanosis, and, if not treated, it can cause death.

Central Nervous System

Dural ectasia, the weakening of the connective tissue of the dural sac encasing the spinal cord, though not life-threatening, can reduce the quality of life for an individual. It can be present for a long time without producing any noticeable symptoms. Symptoms that can occur are lower back pain, leg pain, abdominal pain, other neurological symptoms in the lower extremities, or headaches. Such symptoms usually diminish when the individual lies flat on his or her back. Other spinal issues associated with Marfan's syndrome include degenerative disk disease, spinal cysts and dysautonomia.

Pathogenesis

Marfan's syndrome is caused by mutations in the *FBN1* gene on chromosome 15, which encodes the glycoprotein fibrillin-1, a component of the extracellular matrix. Fibrillin-1 protein is essential for the proper formation of the extracellular matrix, including the biogenesis and maintenance of elastic fibers. The extracellular matrix is critical for both the structural integrity of connective tissue, but also serves as a reservoir for growth factors. Elastin fibers are found throughout the body, but are particularly abundant in the aorta, ligaments and the ciliary zonules of the eye; consequently, these areas are among the worst affected.

Transforming growth factor beta (TGFâ) plays an important role in Marfan's syndrome. Fibrillin-1 directly binds a latent form of TGFâ, keeping it sequestered and unable to exert its biological activity. The simplest model of Marfan's syndrome suggests reduced levels of fibrillin-1 allow TGFâ levels to rise due to inadequate sequestration. Although it is not proven how elevated TGFâ levels are responsible for the specific pathology seen with the disease, an inflammatory reaction releasing proteases that slowly degrade the elastin fibers and other components of the extracellular matrix is known to occur. The importance of the TGFâ pathway was confirmed with the discovery of the similar Loeys-Dietz syndrome involving the *TGFâR2* gene on chromosome 3, a receptor protein of TGFâ. Marfan's syndrome has often been confused with Loeys-Dietz syndrome, because of the considerable clinical overlap between the two pathologies.

Diagnosis

Diagnostic criteria of Marfan's syndrome were agreed upon internationally in 1996. A diagnosis of Marfan's syndrome is based on family history and a combination of major and minor indicators of the disorder, for example: four skeletal signs with one or more signs in another body system such as ocular and cardiovascular in one individual.

Revised Ghent Nosology

According to the US National Marfan Foundation, in 2010 the Ghent Nosology was revised, and new diagnostic criteria superseded the previous agreement made in 1996.

The seven new criteria can lead to a diagnosis.

In the absence of a family history of MFS:

1. Aortic root Z-score >/=2 AND ectopia lentis
2. Aortic root Z-score >/=2 AND an FBN1 mutation
3. Aortic root Z-score >/=2 AND a systemic score >7 points
4. Ectopia lentis AND an FBN1 mutation with known aortic pathology

In the presence of a family history of MFS (as defined above):

1. Ectopia lentis
2. Systemic score >/=7
3. Aortic root Z-score >/=2

Points for systemic score

Wrist AND thumb sign = 3 (wrist OR thumb sign = 1)

Pectus carinatum deformity = 2 (pectus excavatum or chest asymmetry = 1)

Hindfoot deformity = 2 (plain pes planus = 1)

Dural ectasia = 2

Protrusio acetabula = 2

Reduced upper segment/lower segment ratio AND increased arm/height AND no severe scoliosis = 1

Scoliosis or thoracolumbar kyphosis = 1

Reduced elbow extension = 1

Facial features (3/5) = 1 (dolichocephaply, enophthalmos, downslanting palpebral fissures, malar hypoplasia, retrognathia)

Skin striae = 1

Myopia >3 diopters = 1

Mitral valve prolapse (1D 4) = 1

Differential diagnosis

Many disorders have the potential to produce the same type of body habitus (i.e. shape) as Marfan syndrome. Distinguishing among these "marfanoid" disorders can be facilitated by genetic testing, and by evaluating signs and symptoms other than body habitus. Among the disorders capable of producing a marfanoid body habitus are:

- Congenital contractural arachnodactyly or Beals syndrome
- Ehlers–Danlos syndrome
- Homocystinuria
- Loeys–Dietz syndrome
- MASS phenotype
- Shprintzen–Goldberg syndrome
- Stickler syndrome
- Multiple endocrine neoplasia, type 2B

Management

There is no cure for Marfan's syndrome, but life expectancy has increased significantly over the last few decades, and clinical trials are underway for a promising new treatment. At present (2011), the syndrome is treated by simply addressing each issue as it arises and, in particular, preventive medication even for young children to slow progression of aortic dilation if such exists.

Regular checkups by a cardiologist are needed to monitor the health of the heart valves and the aorta. The goal of treatment is to slow the progression of aortic dilation and damage to heart valves by eliminating arrythmias, minimizing the heart rate, and minimizing blood pressure. Beta blockers have been used to control arrythmias and slow the heart rate. Other medications might be needed to further minimize blood pressure without slowing the heart rate, such as ACE inhibitors and angiotensin II receptor antagonists. If the dilation of the aorta progresses to a significant diameter aneurysm, causes a dissection or a rupture, or leads to failure of the aortic or other valve, then surgery (possibly a composite aortic valve graft or valve-sparing aortic root replacement) becomes necessary. Although aortic graft surgery (or any vascular surgery) is a serious undertaking it is generally successful if undertaken on an elective basis. Surgery in the setting of acute aortic dissection or rupture is considerably more problematic. Elective aortic valve/graft surgery is usually considered when aortic root diameter reaches 50 millimeters (2.0 inches), but each case needs to be specifically evaluated by a qualified cardiologist. New valve-sparing surgical techniques are becoming more common.

The skeletal and ocular manifestations of Marfan's syndrome can also be serious, although not life-threatening. These symptoms are usually treated in the typical manner for the appropriate condition, such as with various kinds of pain medication or muscle relaxants. It is also common for patients to receive treatment from a physiotherapist, using TENS therapy, ultrasound and skeletal adjustment. This can also affect height, arm length, and life span. A physiotherapist can also help improve function and prevent injuries in individuals with Marfan's. The Nuss procedure is now being offered to people with Marfan's syndrome to correct 'sunken chest' or (pectus excavatum). Because Marfan's syndrome may cause asymptomatic spinal abnormalities, any spinal surgery contemplated on a Marfan patient should only follow detailed imaging and careful surgical planning, regardless of the indication for surgery.

Treatment of a spontaneous pneumothorax is dependent on the volume of air in the pleural space and the natural progression of the individual's condition. A small pneumothorax might resolve without active treatment in one

to two weeks. Recurrent pneumothoraces might require chest surgery. Moderately sized pneumothoraces might need chest drain management for several days in a hospital. Large pneumothoraces are likely to be medical emergencies requiring emergency decompression.

Research in laboratory mice has suggested the angiotensin II receptor antagonist losartan, which appears to block TGF-beta activity, can slow or halt the formation of aortic aneurysms in Marfan syndrome. A large clinical trial sponsored by the National Institutes of Health comparing the effects of losartan and atenolol on the aortas of Marfan patients was scheduled to begin in early 2007, coordinated by Johns Hopkins.

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Three Stars of Indian Paediatrics

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Abstract

The pionear of Indian paediatrics are not in lime light unfortunately, they have contributed not only in Indian Paediatrics but also at International level. This is the tribute to this great Indian Paediatricians.

Key words: Indian Paediatrics; Dr. George Cohelo; Dr. Professor S.T.Achar; Dr. K.C.Chaudhary.

Dr. George Cohelo

In 1928 he made the beginning of Paediatrics in Mumbai and became the Superintendent of the B.J. (Bai Jerbai Wadia) Hospital for Children - the first children's hospital in India. He remained the head of the department of Pediatrics till 1953. (1) From 1955 to 1971 he was Editor of JAPI (The Journal of the Association of Physicians of India) (2) During period of 1967 to 1969 he was Head of the Departments at L.T.M.C. Medical College, Sion, and Mumbai. He established the Paediatrics in the western part of the country. He was of the opinion that the policies regarding teaching, service and research in Paediatrics should be decided by Paediatricians only.

In 1950 he started the 'Association of Pediatricians of India' (API). From 1952 to 1959 he was editor of the 'Indian Journal of Child Health'.

In 1944 under his guidance the postgraduate diploma in child health was started at Bai Jeerbai Wadia hospital for children. Many of the senior pediatricians of today in the country worked at the BJ Hospital for children under the leadership of Dr George Coelho. (3)He decided to establish the Annual Conference of Association of Pediatricians of India in collaboration with various specialty organizations like Cardiologists, Neurologists, Hematologists and other internal medical disciplines.

He is well known as the 'Father of Paediatrics in India'.

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Professor Dr.S.T. Achar

M.D, FRCP(C), FAMS

(1902-1968)

Dr. S. T. Achar was born on 11th sep. 1902 and retired from service on 10 sep. 1965. (1)(5) He was the pioneer in planning and establisting a separate pediatric facility for the children Institute of Child Health' at Egmore, Madras. This hospital, referred to as the 'Egmore model' has become a model for other public pediatric facilities in India. In 1948 he was the first Professor of Paediatrics in India at the Madras Medical College and first Indian to have a FRCP in Paediatrics from Canada. (4)

His main published work was on paediatrics nutritional problem, childhood cirrhosis & diarrheal disorders which earned him nationwide recognition & international reputation. He was elected as a fellow of Indian academy of medical sciences, honorary fellowship of French, British & American paediatrics society and American academy of paediatrics. He served as a member of scientific advisory board of Indian council medical research for two terms and was the chairman, Nutritional advisory board of Govt. of India after his retirement. He served on health panel of commission & was largely responsible for idea & planning of new children hospital in Egmore, Madras. He was visiting lecturer in various countries & member of International Paediatrics Congress. His knowledge & guidance steered the development of paediatrics in this country.

He passed away June 1968. In memmory of Dr. Achar, a Gold Medal for Social Pediatrics is awarded every year at National Conference of Indian Academy of Paediatrics.(1)

In 1933 he founded the first independent paediatric journal namely, the *Indian Journal of Pediatrics*, in Calcutta. He also started the

Dr. K.C. Chaudhari

Indian Paediatric Society in1948. By 1958 Indian Paediatric Society had organized nine National Pediatric Conferences in different cities in India.He was the first person to highlight the problems of children from east side of India-Calcutta. He was the founder of institute of child health in Calcutta.

He thought that the paediatricians of India should have a separate independent annual conference of their own. He felt that conducting a separate annual meeting of paediatricians would help establish pediatrics as an independent discipline. (1)

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