The Child and newborn

West Bengal Academy of Pediatrics

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The Child and Newborn

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Editorial

As we turn the page to a new volume of Child and Newborn it is a proud and fulfilling moment to share that the journal enters its third uninterrupted year of publication. What began as a humble initiative has now grown into a consistent academic platform—thanks entirely to the collective effort, enthusiasm, and support of every member of the West Bengal Academy of Pediatrics (WBAP). This achievement belongs to all of you.

Our vision for Child and Newborn is ambitious yet clear: we aspire to see the journal indexed and widely recognized. With continued academic contributions from our members, and with your encouragement, we are steadily progressing toward that goal. We believe that with your trust and active involvement, this milestone is well within reach.

These are exciting times for pediatricians across the state. Our academic calendar is vibrant and enriching, filled with diverse opportunities for learning and collaboration. From successful annual conferences hosted by various district branches to impactful sub-specialty chapter meetings, the academic spirit is thriving.

Adding to this momentum is our active participation in the national “Bye Bye Anaemia” campaign. Together with the rest of India, we are committed to reducing the burden of childhood anemia—a cause that directly reflects our dedication to improving child health outcomes.

All these efforts are building up to a momentous occasion—PEDICON 2026which returns to Kolkata after a long hiatus. It is not just an event; it is an opportunity for us to showcase our unity, innovation, and excellence. Let’s come together to make it a PEDICON that will be remembered across the country.

As we step into this promising new year, I wish you all a fulfilling journey of learning, sharing, and meaningful academic endeavors.

Warm regards and best wishes

Prof .(Dr.)Kaustav Nayek
Editor in Chief
Child and Newborn

President's Address

Greetings from the desk of President of West Bengal Academy of Pediatrics.

Dear Colleagues, Friends, and Members of the Pediatric Community,

It is with deep humility and great enthusiasm that I address you as the President of our esteemed Pediatric society, WBAP. As we continue our shared mission of improving the health and well-being of children, I am reminded daily of the profound responsibility and privilege we bear as pediatricians, researchers, and policy makers.

This year, our focus is on equity, innovation, and collaboration. We faced unprecedented challenges in bringing out our beloved journal The Child and Newborn on regular basis . Yet, for last 2 years, it became possible by the energetic Editorial Team to publish this journal with quality articles regularly.

One of our key goals is to strengthen interdisciplinary partnerships, ensuring that pediatric care is holistic and inclusive. We are also committed to expanding mentorship and professional development programs for young pediatricians, fostering a new generation of compassionate and skilled leaders.

I am especially proud of the initiatives we've launched to support community-based care, harness digital health technologies, and promote evidence-based practices through robust, collaborative research. Our journal remains at the heart of these efforts, serving as a vital platform for knowledge sharing and innovation.

As we look ahead, I invite each of you to engage, contribute, and lead. Together, we can create a healthier, more equitable future for all children.

If we can sail through all the odds, I am sure that our Child and Newborn journal would be an indexed journal very soon.

Thank you for your dedication, passion, and partnership.

Warm regards,

Prof Swapan Kumar Ray
President 2025, WBAP

Joint Statement on Triple Elimination of Vertical Transmission of HIV, Syphilis & Hepatitis B (TEVTHSH) Initiative of West Bengal: Indian Public Health Association (IPHA), West Bengal Academy of Pediatrics (WBAP), Neonatology Society of West Bengal(NSWB), Bengal Obstetricand Gynecological Society (BOGS), Associationof Physician of India (API), Indian Association of Dermatologists, Venereologists and Leprologists West Bengal (IADVL WB)

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Preamble

We, the professional associations as advocates and contributors to health and development of the people in India, have come together for promoting elimination of vertical transmission of diseases, aligning with the theme of World Health Day 2025 "Healthy Beginnings,HopefulFutures," focusing on improving maternal and newborn health and survival. Achieving the triple elimination of vertical transmission of HIV, Syphilis and Hepatitis B is now a global health priority. Infected infants in all three conditions are prone to higher rates of mortality and morbidity, and experiencelifelong consequences. Congenital syphilis continues to be the second leading cause of stillbirth.Earlyexposurevia verticaltransmissionorduringearlychildhoodisa major factor in the overall disease burden of HBV. In the absence of preventive measures for mothers and children, the risk of vertical transmission ranges from 70-90% in cases of high maternal viral load. In India, presence of a robust RMNCH+A framework and disease-specific programs (NACP and NVHCP), present opportunities for the implementation of a Triple EMTCT strategy which leverages the capabilities of the current programs and achieves the desired impact with greater efficacy and efficiency. This joint statement proclaims our collective commitment to work with the whole society including the affected community toward holistic healthy development and well-being of mother & newborn of our country; every child should be given the best chance to start a healthy life, born free of preventable infections.

Context

West Bengal is the first state in the country to undertake TEVTHSH initiative with the aim of elimination vertical transmission of HIV, Syphilis and Hepatitis B by 2026. Rationale of Triple Elimination of HIV Syphilis and Hepatitis B lies in the similarity of modes of transmission, the prolonged period of silence and latency in

the infected mothers with the potential to cause life-long consequences and significant morbidity and mortality in the mother-baby pair. Antenatal care, including regular check-up, happens tobe anestablished gateway to screening, identification,prophylaxisand management for all the three infections. The state of West Bengal has a strong track record in service delivery for HIV, Syphilis, and Hepatitis programs. The state has made exceptional progress in reaching out to high-risk groups, such as those in closed settings, engaging with the private sector and professional bodies. Moreover, West Bengal has established effective coordination mechanisms between different health programs. Currently, West Bengal's overall antenatal care performances are at par with or surpass the national averages, which have paved the way for undertaking TEVTHSH initiative.

The build-up summary

The blueprintof theinterventionwasforged inmid-2023throughconsultations among the officials of the State Task Force (STF) and Technical Advisory group (TAG), with the former having key-officials of all relevant program divisions and the later a multidisciplinary group with the coherent technical experts. Field-level actionstowards the initiative began with an elaborate pre-pilot situational assessment in 4 select districts (Sep-2023) that culminated in the development of a minutely workedout contextual intervention package. The explicit Declaration of Intent of the state was made through the Kolkata Commitment in a mega Consultation in December 2023. A pilot was thereafter implemented from April-2024. Based on the encouraging learning from the pilot, the State has now moved to a State-wide scale-up of the TE initiative from early 2025. William J Clinton Foundation (WJCF), all major ProfessionalAcademic Organizations, viz., FOGSI, API, IAP, IPHA, IADVL, IAPSM, NNF, CINI India and WHO have been partners in this journey. Earliest detections in pregnancy andleakprooflinkagetocare,topreventverticaltransmission,aretheprime objectives. A person-centered systems approach has been at the core of the initiative beginning with obstetric care to infection specific care of mother to care and follow up of the exposed baby.

TEVTHSH Framework

A detailed cross-sectional assessment across 150 relevant service delivery points in the 4 selected districts was conducted during pre-pilot situational assessment, which also factored around 40 infected beneficiaries receiving services. The areas of improvement emerging from the assessment were: standardized protocol for referral, linkage, treatment and care; standardized inventory and commodity management; strengthened recording and reporting with back-and-forth communication channels; governance system for sustained accountability. A need-based intervention package with the following key considerations was developed, based on the findings:

(a) Development of SOP module with standardized protocol for screening, referral, linkage, treatment and care of pregnant women and baby, till the age of 18 months (for both public and private health facilities).

(b) Guideline for behavioral risk-profiling of the screened negative pregnant woman has been introduced to ensure necessary retesting at later stages of pregnancy.

(c) Maternal syphilis management has been decentralized to Block-level along with availability of Benzathine Penicillin-G (BPG). Pregnant woman screening reactive for syphilis are entitled to receive all 3 weekly doses of BPG.

(d) Decentralization of delivery of HIV and HBV positive mother at block level.

(e) Ensuring timely Hepatitis B Birth Dose (HBBD) vaccine administration to all newborns and both HBBD & Hepatitis BImmunoglobulin(HBIG) administration to the exposed infant on time. Private facilities may avail HBBD and HBIG on request from the respectiveDistrict Health Authorities for the beneficiarieswho are in need and administer at free of cost. This activity needs extensive promotion in private sector and involvement of more representatives from private facilities. Many private facilities are still not administering HBBD to newborn.

(f) Relooking existing synergy between the relevant program divisions and strengthening coordination among them.

(g) Establishing coordination between Labour room, SNCU, ICTC, ARTC, VHTC and DSRC to ensure ensured appropriate management of HIV, Syphilis and HepatitisB positive mother and exposed infant.

(h) RevampingGovernancestructurewithsetkeyrolesand responsibilities.

(i) Guidance on key commodities, such as testing, prophylactic, treatment commodities.

(j) Development and distribution of IEC materials on TEVTHSH for both for both public and private health facilities.

(k) Establishing robust supervision and monitoring framework along with a dedicated web portal for data reporting (capturing information from obstetric care to disease specific care of mother and appropriate management and follow up of the exposed infant).

Driven by significant progress and learning from the pilot, the Government of West Bengal has rolled out the EVTHSH initiative across the entire State in April 2025. Accordingly, capacity building for rest of the districts to equip district workforce to undertake EVTHSH initiative is ongoing. Concerned state program divisions are ensuring availability of key commodities. Focus is also on increased engagement with private health facilities and professional bodies working in the coherent fields.

Callto Action

We, the related professional associations, commit ourselves and call upon other stakeholders to support and facilitate, without delay, the following actions to ensure that all pregnant women and newborn, especially HIV, Syphilis and Hepatitis B positive mothers and their exposed infants, receive appropriate preventive and therapeutic services.

(a) Harmonization of multisectoral policies and strategies to eliminate vertical transmission of HIV, Syphilis and Hepatitis B.

(b) ActiveinvolvementandpartnershipofHealthcarefacilitiesandProvidersengaged in the Healthcare Delivery in the Private Sector.

(c) Establishment of linkage between private practitioners and Health Authorities for better tracking of beneficiaries and ensuring appropriate management on time.

(d) Increase awareness and engagement of all relevant stakeholders, community, governmentfunctionaries,politicalandreligiousleaders,civilsocietyorganizations and international organizations.

(e) Capacity building and training of members of the professional associations to ensure appropriate management of pregnant women and exposed infants as per TEVTHSH protocol.

(f) Strengtheningofmonitoringandevaluationsystemthroughcollectionandanalysis of good qualitydata which can contribute todecision-making process and program management.

(g) Dialogue and cooperation with policy makers and programmes heads of National and State level regarding seamless implementation of effective TEVTHSH.

(h) Role of professional associations are pivotal in community awareness generation regarding TEVTHSH. Their collective action can also promote inclusivity and reduce remnants of social stigma.

(i) Including TEVTHSH as agenda in academic discussions/conferences ofprofessional associations for its widespread circulation among the stakeholders.

(j) Our collective aim is to ensure appropriate counseling, screening and management of all pregnant women and newborn to enable them to live life and thrive with good health, well-being, and dignity.

Way Forward

On this day of World Health Day, 7th April 2025, let us unite and take a pledge to eliminate the vertical transmissionof HIV, Syphilis and Hepatitis B in the state of West Bengal. We can collaborate to implement standardized screening and management protocol in West Bengal. We can conduct capacity building programmes to enhance healthcare worker's skill in early diagnosis, prenatal and postnatal care, newborn care and preventive approaches. Thus, together we advocate for community awareness initiative to promote "Healthy Beginning, Hopeful Future."

We, the professional associations, affirm our commitment as partners to the West Bengal initiative of Triple Elimination of Vertical Transmission of HIV, Syphilis & Hepatitis B by 2026.

Published on WORLDH EALTH DAY 2025

Date: 07.04.2025 Place: Kolkata

Allergic Broncho-Pulmonary Aspergillosis (ABPA) and Childhood Asthma – A management Dilemma

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Abstract:

Treatment of ABPA (Allergic Broncho-Pulmonary Aspergillosis) in children in absence of an underlying lung disease can be challenging to the clinicians. An aggressive management may be indicated to prevent long term damage whereas the treatment options do come with significant side effects. If started on treatment, there is a need for close monitoring and increased time spent in the hospital hampering education and holistic growth and development of a child. Hence, a decision to treat needs to be taken with caution. We discuss a teenager who attended paediatric respiratory clinic with wet cough and asthma and was diagnosed with ABPA serologically. In absence of radiological findings, patient was not actively treated for ABPA. We report outcome at two years.

Key words : Asthma, ABPA, Children

Conflict of interest and consent

Author has no conflict of interest. A verbal consent was taken from the parents by the responsible clinician (author) to use relevant investigations and case history anonymously (no patient identifiable data has been used).

Background

Allergic bronchopulmonary aspergillosis (ABPA) is a common allergic respiratory mycosis in patients with asthma and cystic fibrosis in adults. It has a global prevalence of 11.3% (95% CI, 8.7-14.2) in adult asthma patients. It is an allergic respiratory mycosis caused by Aspergillus fumigatus. In the paediatric world a diagnosis of ABPA without a background of Cystic Fibrosis or any other interstitial lung disease is rare. The experience of physicians in the treatment of these patients are minimal. Aggressive treatment is not risk free. On the other hand if left untreated this may cause long term damage to the lungs. Hence, there is a need to cohort these patients and do subsequent follow ups into their adult life.

Case presentation

A fifteen and half year old girl was first seen in a respiratory clinic with poorly controlled asthma. Patient was known to the general paediatrician for a long time as she was getting treated with a diagnosis of asthma for a very long time. She mentioned that her asthma used to be well controlled on inhaled steroid and long acting beta agonist combination (Fluticasone 50 mcg + Salmeterol 25 mcg, 2 puffs twice a day). However in the last 4 years her paediatrician was struggling to manage her asthma. Her inhaler was changed to a higher dose (Fluticasone 125 mcg + Salmeterol 25 mcg, 2 puffs twice a day) with no added advantage. With a desperate attempt to help her, Montelukast was added which didn't help her either. She reported that her symptoms were mainly wet cough especially in the morning. No significant wheeze or breathing difficulties were reported but the cough sometimes sounded like smokers cough. Salbutamol was not helping either. There was no history of smoking or exposure to cigarette smoke. A skin prick test done in the past showed positive reaction to house dust mite, aspergillus, grass pollen and tree pollen. Further history was taken regarding any environmental changes that happened 4 years back. She mentioned that the only change was that she started horse riding.

Investigations and clinical course

Initially a chest X-ray was done which was unremarkable apart from hyper expansion and some

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		Best	SR	% Pred	Pred	Pred LL	Post	% Change
Level date		27.10.22					27.10.22	
Level time		10:04					10:34	
Substance							Salbutamol	
Dose							400 mcg	
FEV1	L	2.62	-1.05	87.7	2.99	2.41	2.85	8.8
VC MAX	L	2.90	-1.08	87.0	3.34	2.68	2.96	1.9
FEV1%M	%	90.32	0.06	100.4	90.00	78.72	96.44	6.8
PEF	L/min	321	-0.58	88.5	363	243	351	9.3
MMEF	L/s	3.63	-0.06	98.7	3.68	2.46	3.56	-2.1
TLCOcSB	mmol/(min*kPa)	7.52	0.82	114.3	6.58	5.10		
KCOc	mmol/(min*kPa*L)	1.88	0.79	112.6	1.67	1.28		
Hb	g(Hb)/dL	13.80						
VA_SB	L	4.01	0.09	101.1	3.96	3.25		
VIN_SB	L	2.83	-1.26	84.9	3.34	2.68		

Pic 1: Lung function

bilateral streaky changes. She described the sputum as brownish and it was sent for extended microbiology screening. Sputum did grow Pseudomonas mendocina sensitive to Amikacin. Though this is usually a commensal (however, isolated growth of one species is not common), she was treated with two weeks course of oral Ciprofloxacin (standard treatment Pseudomonas in sputum in symptomatic patient). Further investigations were requested at this time.

Lung function was normal without any significant obstruction and reversibility to bronchodilator with a FeNO of 7 ppb (normal <35 ppb). (Pic 1) Skin prick test was repeated at the same time. (Pic 2).

With the history of exposure to damp environment of stables for horse riding, production ofbrown sputum, very high IgE and SPT positive to Aspergillus, standard blood tests for ABPA i.e.IgE to Aspergillus, IgG to Aspergillus (precipitins) and total IgE were checked and they all came back very high. This raised the question whether an interstitial lung disease was missed. This patient was born before the new born screening was introduced in England. Hence a sweat test was done to rule out cystic fibrosis which was negative. Patient being white British, genetic testing was not done as the sweat test was negative unequivocally.

Differential diagnosis

We looked at two guidelines for diagnosis of ABPA widely used:

Negative control	Negative
Positive control	4 x 4 mm
Early tree (Betulacee mix)	10 x 9 mm
Birch tree	3 x 3 mm
Plane tree	12 x 6 mm
Mixed grass	9 x 9 mm
Timothy grass	4 x 4 mm
Ragweed	4 x 3 mm
Mugwort	4 x 5 mm
Alternaria	5 x 5 mm
Aspergillus	7 x 5 mm
Cladosporium	4 x 5 mm
D Farinae	8 x 9 mm
D Pteronyssinus	7 x 10 mm
Cat	5 x 5 mm
Dog	5 x 5 mm
Horse	6 x 5 mm
Rabbit	6 x 5 mm

Pic 2: Skin prick test repeated at the same time

1) Rosenberg-Patterson criteria: (ref 1)

It has eight major and three minor criteria

Major criteria

1. Asthma
2. Presence of transient pulmonary infiltrates (fleeting shadows)
3. Immediate cutaneous reactivity to Af (A. fumigatus)
4. Elevated total serum IgE
5. Precipitating antibodies against Af
6. Peripheral blood eosinophilia
7. Elevated serum IgE and IgG to Af

8. Central/proximal bronchiectasis with normal tapering of distal bronchi

Minor criteria1.

Expectoration of golden brownish sputum plugs2. Positive sputum culture for Aspergillus species3. Late (Arthus-type) skin reactivity to Af

2) Criteria proposed by ISHAM working group (ref 2):

Predisposing conditions1.

Bronchial asthma2. Cystic fibrosis

Obligatory criteria (both should be present)

1. Type I - positive Aspergillus skin test (immediate cutaneous hypersensitivity to Aspergillus antigen) or elevated IgE levels against Af
2. Elevated total IgE levels (greater than 1000 IU/mL)

Other criteria (at least two of three)

1. Presence of precipitating or IgG antibodies against Af in serum
2. Radiographic pulmonary opacities consistent with ABPA
3. Total eosinophil count over 500 cells/microliter in steroid naïve patients(If the patient meets all the other criteria, an IgE value less than 1000 IU/mL may be acceptable)

Our patient did qualify for a serological diagnosis of ABPA (without any radiological feature). However, in absence of a chronic lung disease like Cystic Fibrosis, ABPA is uncommon in children. ABPA and asthma are more associated in the adult population. Hence there was a dilemmawhether to treat or not with long term steroid and antifungal. Long term treatment with oral steroid in paediatric population needs a confirmed diagnosis as this has significant side effects. There was no bronchiectasis i.e. no radiological diagnosis of ABPA and apart from one isolation of Pseudomonas species, all other sputum samples was negative for any significant pathogen (isolated only normal respiratory flora). A normal lung function with no significant reversibility and normal FeNO ruled out severe asthma with fungal sensitivity (SAFS).

Treatment

Patient was discussed in the team involving three respiratory paediatricians. A wait and observe approach was taken while the patient was advised against horse riding, hence exposure to Aspergillus in the stables was stopped. Asthma management continued with inhaled steroid and long term beta agonist combination. No oral steroid or anti-fungal was given. Subsequent blood tests showed a declining ABPA markers (table 1). At the age of sixteen and half patient was transitioned to the adult respiratory team.

Outcome and Follow up

With the adult respiratory specialist team, the patient was changed to Fostair 100/6 (Beclometasone and Formoterol) as the serological markers were improving. Diagnosis was atopic asthma with significant sensitizations to aero allergens (pic 4) including multiple fungal aeroallergens. After being on the new inhaler for nearly a year, she has now been changed to AIR therapy with the same inhaler (anti-inflammatory reliever therapy) and continues to make progress.

Table 1. Subsequent blood tests showed a declining ABPA markers

Date	Total IgE (normal 0-75)	Aspergillus FumigatusIgE (normal 0-0.35)	Aspergillus precipitins (IgG) (normal 0-39.9)	Eosinophil 10*9/L Normal 0-0.5
10/10/2022	3185 kU/ L			0.2
9/1/2023	1575 kU/L	18.2 kUA/L	178 mgA/L	
22/2/2023	1752 kU/L	17 kUA/L	168 mgA/L	
28/6/2023	1304 kU/L	10.5	154	
9/1/2024	1269 kU/L	7.25 kUA/L		0.21

ALLERGENS	
Dermatophagoides Pteronyssinus ...	49.70
Cat Dander	20.40
Horse Dander "Equus caballus"	11.30
Dog Dander "Canis familiaris"	8.04
Rabbit Epithelium "Oryctolagus cu...	>100.00
Penicillium chrysogenum "P.notat...	1.02
Cladosporium herbarum "Hormod...	57.70
Aspergillus fumigatus (IgE)	7.25
Alternaria alternata	1.59
Grass Pollen mix GX1 (G3, G4, G5,...	>100.00
Mould mix MX2 (M1,M2,M3,M5,M6,...	19.20
Tree pollen mix TX1 (T1, T3, T7, T8...	38.70

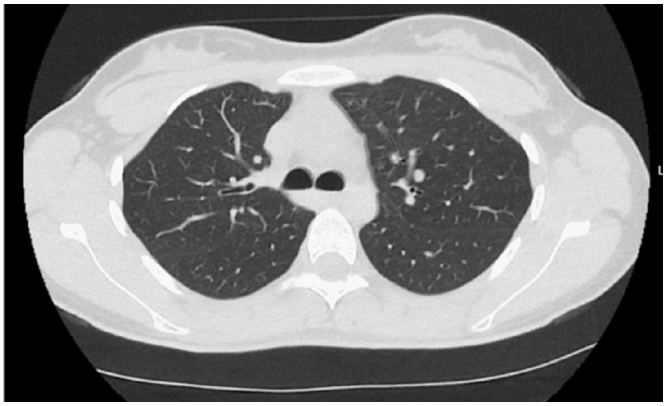
Discussion

Treatment for ABPA is long term with significant impact on overall health. One way of treating is with oral steroid for 5-6 months along with anti-fungal therapy. This has got significant side effects and can be detrimental to a child as it involves prolonged use of steroid. Other treatment available is anti IgE (Omalizumab) which is an injectable medicine needing close monitoring, intervention and multiple attendance to hospital. Hence a more conservative approach was taken. Our advantage was the absence of radiological changes suggestive of ABPA. In medium term this has worked and the patient is getting better with a reduction in treatment load. Whether she will need treatment at an older age, we do not know.

Picture 3: CT chest

Report: There is mild airways thickening, likely to be inflammatory. There is no evidence of bronchiectasis. No evidence of pulmonary fibrosis. No parenchymal lung abnormality identified.

No mediastinal or hilar adenopathy.



Pic 3: **CT chest**

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Research Paper submitted for Dr. James Flett Endowment Award in "Social and Preventive Pediatrics".

A Study on the Prevalence & Correlates of Nicotine Dependence & Its Relation with Quitting among School-going Children of Burdwan Town, West Bengal

Dr Kamirul Islam
Pediatrician , Katwa Sub divisional Hospital

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Introduction:

Substance abuse is becoming a public health concern in every corner of the world and India is of no exception.[1] Excessive use of tobacco both in the form of smoking & smokeless tobacco is an example of modern epidemic and also known as 'the brown plague'.[2] WHO estimated that tobacco use will cease one billion lives in the 21st century & 80% of that will occur in developing countries.[3] Decreased awareness among people, risky mind set up of adolescents, poor literacy and repeated advertisements by tobacco companies in mass media- all are responsible for prevalence of tobacco use. [4, 5] All these lead to substance abuse among adolescents leading to economic burden due to health problem. Tobacco use is also prevalent school going children of India. [6, 7]

Use of tobacco causes temporarily pleasing effect in brain predominantly by altering the mesolimbic pathway. [8] Nicotine is the chief chemical in tobacco which causes dependence- both physical & psychological. The same also stands true for smokeless forms of tobacco. Cotinine is a metabolite of nicotine which is measured in serum/ saliva/urine to find the level of nicotine dependence of a person. [9, 10] But this test is difficult to perform and costly. Hence many questionnaires were developed which act as a surrogate marker of nicotine dependence. One such questionnaire is Fagerström Test for Nicotine Dependence (FTND). The questionnaire was used in different parts of world & its reliability is confirmed in different population. [9, 10] This degree of nicotine dependence will further be helpful in determining suitable plans for cessation of tobacco use (counseling/ pharmaco-therapy/ both). [8]

Many studies had been carried out previously to find the prevalence of tobacco use among Indians. But detail information regarding prevalence of different levels of nicotine dependence in school-going children and is not available in Indian context. In this background this study was conducted among adolescent tobacco users of Burdwan town to find out prevalence of different levels of nicotine dependence among adolescent tobacco users & factors responsible for it. Another aim of the study was to find out the relationship between nicotine dependence and quitting tobacco, if any.

Materials & Methods:

A school-based observational cross-sectional study was conducted in Burdwan town between January 2013 to December 2017 after taking permission from Institutional Ethics Committee [vide memo no BMC/ PG/2725]. Informed written consents were taken from the participants/ their legal guardians, as applicable. A pre-designed pre-tested semi-structured schedule was used for collection of data by school visit. All the current tobacco users constituted our study population & complete enumeration method is thus followed. Total number of school-going (secondary and higher secondary) students in the study area was 16332. [Source-District Board of Education] Prevalence of tobacco use among Indian school-children is 14.4%. [Source-Global tobacco survey] So, approximate number of tobacco-users is 2352. But a total of 2481 tobacco users were identified & 23 were excluded from the study (1-seriously ill, 16-absent despite three visits, 6-consent was not available /denied to answer questions). So, 2458 tobacco users were interviewed

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and they constitute our study population. The interviewer, interviewee & the data entry operators-all of them were not aware of the purpose of study.

Operational definitions:

People who were smoking at the time of study & had smoked>100 cigarettes in their lifetime were defined as current smokers. [11] Current smokeless tobacco users were defined as people either chewing or snuffing tobacco at the time of study and either had snuffed or chewed tobacco more than 20 times in their lifetime. [12] Cigarettes, beedi, cigars, pipes etc were considered as smoking products. Smokeless tobacco products include chewing tobacco & moist/dry snuff. Low, medium & high level of nicotine dependence were defined as FTND score <4, 4-6 & >6, respectively. [9, 10] Who were registered student of school at the time of study was considered as the school children. Abstinence from tobacco for one year was considered as quitting.

Statistical analysis:

All the collected data were entered into Microsoft Excel Worksheet (Microsoft, Redwoods, WA, USA) after double checking. Categorical & continuous data were expressed in proportion & mean values, respectively. Kolmogorov Smirnov test revealed that the data were normally distributed (as n>2000) . Significance of association between two attributes in contingency table was assessed by Pearson's chi-square ($\chi 2$) test. Significance of difference between two means was tested by Student's independent t-test (unpaired), while one-way ANOVA (Analysis of variance) was used for comparing >2 means. Categorical variables were coded. Degree and direction of relationship between FTND score and different study variables was computed by Pearson's product moment correlation coefficient (r). Significantly correlated variables were further considered for multivariable linear regression analysis taking FTND score as dependent variable. Another binary logistic regression model was created taking quitting as an outcome variable. Adjusted odds ratio was calculated with 95% confidence interval. P<0.01 was considered as statistically significant. All the data were rounded off upto one decimal point. All the statistical analysis was done by SPSS version 19.0 (Statistical Package for the Social Sciences Inc, Chicago, IL, USA).

Results:

Mean age of the study population is 16.9(2.5) years with majority being >15 years of age (60.8%) Out of the 2458 tobacco users, majority was male (94.2%), Hindu (85.4%) & belonged to joint family (62.6%) & lower SES (51.3%). 38% of the tobacco-users were exclusively smokers, 37 % were using smokeless tobacco exclusively & 25% were using both. 32.1%, 32.6% and 35.3% of tobacco-users belonged to high, medium and low dependence group, respectively. Mean FTND score of all tobacco users was 4.2(2.1). After repeated counseling and pharmacotherapy by the adolescent friendly health clinic, 652 tobacco users (26.5%) were able to quit tobacco.

High Nicotine Dependence was maximally observed among users who were >15 year of age (42.2%), males (33.4%), Hindu (33.8%), who belonged to nuclear family (48.6%), middle SES (35.2%),started using tobacco <10 years (53.6%), using tobacco for >5 years (50.0%), got <40% marks in last test (41.6%), belonged to problem family (42.5%), daily tobacco users (41.1%) & were not aware of the injurious effect of tobacco (49.6%). These differences were found to be significant by chi-square test. Mean FTND score was significantly higher among school-children aged >15 years, males, Hindu, tobacco-users of nuclear family, who belonged to upper socioeconomic status, who started using tobacco at the age of <10 years, using tobacco for =5 years, got <40% marks in last examination, belonged to problem family, daily users and not aware of the injurious effect of tobacco to health. [Table I]

There was statistically significant positive correlation of FTND score with age (r=0.65), male sex (r=0.17), Hindu religion (r=0.23), nuclear family (r=0.34), increase in socioeconomic status (r=0.53), duration of tobacco use (r=0.76), working (r=0.421) & daily tobacco use (r=0.78) implying that increase in this variables will increase the FTND score. FTND score is significantly and negatively correlated with starting age of tobacco use (r=-0.59), marks in last examination (r=-0.26), & knowledge of injurious effect of tobacco to health (r=-0.52) implying that increase in this variables will decrease the FTND score.

A linear regression model was generated using FTND score as the dependent variable. Significantly

Table I. Table I: Distribution of study population according to level of nicotine dependence & different variables (n=2458)

Variables	Low N (%)	Medium N (%)	High N(%)	FTND score Mean(SD)	Significance
Age					
=15 years	612(63.5)	195(20.3)	157(16.2)	2.3(1.4)	t=64.2, P<0.001
>15 years	257 (17.2)	606 (40.6)	631 (42.2)	5.7(1.2)	$\chi 2$ =552.4, P<0.001
Sex					
Male	775 (33.5)	766 (33.1)	774 (33.4)	5.3(2.1)	t=19.3, P<0.001
Female	94(65.7)	35 (24.5)	14(9.8)	1.9 (0.8)	$\chi 2$=66.2, P<0.001
Religion					
Hindu	794 (37.8)	595 (28.4)	710 (33.8)	4.2 (2.4)	t=17.6, P<0.001
Others	75 (20.9)	206 (57.4)	78 (21.7)	1.9(1.4)	$\chi 2$=118.2, P<0.001
Family type					
Nuclear	310(33.7)	163 (17.7)	447 (48.6)	5.7(2.2)	t=32.7, P<0.001
Joint	559 (36.3)	638 (41.5)	341 (22.2)	2.7(2.2)	$\chi 2$=226.2, P<0.001
Socioeconomic Status					
Upper	143 (31.3)	159 (34.8)	155 (33.9)	5.7(1.3)	F=104.8, P<0.001
Middle	124 (16.8)	355 (48.0)	261 (35.2)	4.9(2.2)	$\chi 2$ =226.5, P<0.001
Lower	602 (47.7)	287 (22.8)	372 (29.5)	4.2(2.0)	
Starting Age					
<10	50 (5.7)	356 (40.7)	469(53.6)	5.8(1.3)	F=887.8, P<0.001
10-12	350 (38.1)	314 (34.2)	255 (27.7)	3.5(2.0)	$\chi 2$ =741.2, P<0.001
>12	469 (70.6)	131 (19.7)	64 (9.7)	2.6(1.1)	
Duration-					
<5 years	583 (41.7)	557 (39.8)	258 (18.5)	2.9(1.6)	t=51.8, P<0.001
=5 years	286 (27.0)	244 (23.0)	530 (50.0)	6.1(1.4)	$\chi 2$=276.4, P<0.001
Marks in the last test-					
>60%	349 (67.2)	110 (21.2)	60 (11.6)	3.2(0.9)	
41-60%	232 (44.1)	154 (29.3)	140 (26.6)	5.1(1.7)	F=698.1, P<0.001
<40%	288 (20.4)	537(38.0)	588 (41.6)	6.3(1.8)	$\chi 2$= 398.7, P<0.001
Family status					
Normal	457 (38.8)	477 (40.5)	245 (20.7)	2.1(2.0)	t=34.7, P<0.001
Problem in Family	412 (32.2)	324 (25.3)	543 (42.5)	4.9(2.0)	$\chi 2$ =140.4, P<0.001
Habit					
Daily	347 (25.0)	470 (33.9)	569 (41.1)	5.7(1.6)	t=66.5, P<0.001
Occasional	522 (48.7)	331 (30.9)	219 (20.4)	1.9(1.1)	$\chi 2$ = 177.6, P<0.001
Knowledge about injurious effect of tobacco					
Not Known	316 (27.6)	261 (22.8)	570 (49.6)	5.2(2.0)	t=12.0, P<0.001
Known	553 (42.2)	540 (41.2)	218 (16.6)	2.1(0.8)	$\chi 2$ = 309.5, P<0.001

correlated variables in the correlation analysis were further considered for regression analysis. Family type, religion & knowledge of injurious effect of tobacco use though significantly correlated with FTND score, become insignificant in the regression model. Our model can correctly predict 84.8% variation of the dependent variable i.e. FTND score. [Table II]

Maximum tobacco users who can quit tobacco successfully belonged to the groups who started tobacco use =10 years (34.8%), using tobacco for <5 years (32.3%), low nicotine dependence, occasional tobacco users (52.6%) and were aware of the injurious effect of tobacco to health (33.8%). Mean FTND score was also significantly lower among the tobacco users who were able to quit tobacco. [Table III]

Quitting is significantly and negatively associated with FTND score (r=-0.73), daily tobacco use (r=-0.65), & duration of tobacco use (r=-0.62) signifying that they were associated with difficulty in quitting. On the other hand quitting is significantly and positively associated with starting age of tobacco use (r=0.39) signifying that when starting age of tobacco use increases quitting become easier. [Table IV] Our model can explain 67.9%-78.5% variability of outcome variable. Out of this, 62.1% is due to FTND score alone.

Discussion

Tobacco use both in the form of smoking & smokeless tobacco ultimately results in large economic burden to the government due to health related issue. Our study was conducted to find out the prevalence of different levels of nicotine dependence among adolescents and its correlates. We found that 35.3%, 32.6% & 32.1% tobacco users belonged to low, medium & high dependence group respectively. High dependence is lower than the findings of Clemente Jiménez et al (3.3%) may be due to higher socioeconomic status of their study population.[13] Saha I et al also noticed a higher prevalence of high nicotine dependence (57.8%) mainly because they included adult tobacco users and nicotine dependence gradually increases with age.[14] Mean FTND score was 4.2(2.1) which is similar to the findings of Fagerström K et al (2.8-4.6) & Jayakrishnan R et al [5.04(5.05)].[15,16] But Saha I et al found a higher score [6.47(2.38)] may be due to inclusion of adult tobacco users.[14]

Similar to the observation of Wu J et al, Jayakrishnan R et al & Saha I et al, we also found a gradual rise of FTND score with increase in age.[14,16,17] Mean FTND score was significantly higher among males [5.3(2.1)], Saha I et al also noticed the same but their finding is not significant (P>0.05).[14] Roberts B et al found that bad socioeconomic status was

Table II. Regression coefficients in multivariable linear regression (enter method) taking FTND score as dependent variable. (n=2458)

Variables	B (95% CI)	SE	Beta	t	Sig
Constant	-1.305 (-2.050- -0.560)	0.380		-3.435	0.001
Age	0.504 (0.158-0.849)	0.176	0.563	2.859	0.004
Sex	1.214(0.880-1.547)	0.170	0.116	7.142	<0.001
Religion	-0.183(-0.391-0.024)	0.106	-0.020	-1.731	0.084
Starting Age	-0.396(-0.743- -0.049)	0.177	-0.324	-2.239	0.002
Duration	0.082(-0.268-0.432)	0.178	0.089	0.458	0.006
Family	-0.127 (-0.258- 0.004)	0.067	-0.023	-1.900	0.058
Socioeconomic Status	0.225 (0.132-0.317)	0.047	0.063	4.758	<0.001
Marks in Exam	-0.256 (-0.342- -0.169)	0.044	-0.075	-5.771	<0.001
Habit	1.425 (1.262-1.588)	0.083	0.292	17.212	<0.001
Effect	0.160(0.153-0.169)	0.074	0.012	0.024	0.02

Table III. Regression coefficients in multivariable linear regression (enter method) taking FTND score as dependent variable. (n=2458)

Variables	Quitted Tobacco No(%)	Not quitted tobacco No(%)	Significance
Starting age			
<10 years	101(11.5)	774 (88.5)	$\chi 2$=118.2
=10 years	551 (34.8)	1032 (65.2)	P<0.001
Duration			
<5 years	452(32.3)	946 (67.7)	$\chi 2$=56.1
=5 years	200 (18.9)	860(81.1)	P<0.001
Nicotine Dependence			
Low	388 (49.2)	400 (50.8)	$\chi 2$=513.0
Medium	256 (32.0)	545 (68.0)	P<0.001
High	8 (0.9)	861 (99.1)	
FTND score	3.1 (1.2)	5.9 (1.5)	Student's t=43.0, P<0.001
Habit			
Daily	88 (6.3)	1298 (93.7)	$\chi 2$=663.7
Occasional	564 (52.6)	508 (47.4)	P<0.001
Knowledge of injurious effect			
Yes	443 (33.8)	868 (66.2)	$\chi 2$=76.1
No	209 (18.2)	938 (81.8)	P<0.001

Table IV. Adjusted odds ratio & correlation coefficient for quitting tobacco (n=2458)

Variables	Correlation coefficient	Adjusted odds ratio (95% CI)
FTND Score	-0.73*	0.31* (0.25-0.37)
Daily use	-0.65*	0.47* (0.44-0.50)
Duration	-0.62*	0.51* (0.47-0.54)
Starting age	0.39	1.73* (1.60-1.76)

**Significant at the 0.01 level*

associated with higher level of nicotine dependence.[18] In contrary we found that prevalence of high nicotine dependence was more among students of middle socioeconomic status and mean FTND score was highest among students of upper socioeconomic status. We observed a gradual decrease in the FTND score with increase in performance in examination. Similarly, Schmidt A et al also noted lower education as a risk factor for higher nicotine dependence.[19] Jayakrishnan R et al & Wu J et al, both noted an increase in the score with increase in education, which is contrary to our findings.[16,17] Similar to the findings of the present study, Roberts B et al, Taioli E et al & Breslau et al also found that initiation of tobacco use at an early age & using tobacco for longer duration were significant risk factors for high level of nicotine dependence. [17,20,21] Similar to the finding of Saha I et al we also observed that FTND score was higher among daily users & who were not aware of the injurious effect of tobacco on health.[14] Our model can correctly predict 84.8% variation of the FTND score, while Saha I et al can only predict 27.3%.[14]

In our study only 652 users successfully quitted tobacco which is lower that other Indian study. Abdolahinia A et al reported that mean age of initiation of tobacco use was higher among quitters. [22]We also noted the similar findings, may be due to less FTND score among the users who started using tobacco late. Similar to Bernstein SL et al, we also noted that FTND score is the single most significant factor responsible for quitting.[23] Islam K et al also noted the same observation among adult tobacco users.[24]

Conclusions

It is clear that tobacco use & dependence to nicotine had its origin at adolescence and hence school children should be targeted and made aware of

injurious effect of tobacco to health. Their parents and teachers should actively be involved. Advertisements against use of tobacco should be done in TV/newspapers/radio to prevent nicotine dependence. “Get ready for plain packaging”- the slogan for world no tobacco day (2016) should be given due importance.13Suitable plans should be developed for those who want to quit tobacco depending on their nicotine dependence.[8] Though this study contains an appropriate sample size, for development of appropriate plans, multi-centric studies should be undertaken. As the study was conducted in urban area, it may not reflect the rural scenario as well.

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A Population Based Model for Congenital Heart Disease- Hridyam: What has been Accomplished and What are the Current Challenges?

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Background:

Congenital Heart disease (CHD) is the most common serious birth defect that affects children and is responsible for more deaths than any other birth defect. Between 8–10 children for 1000 live births have CHD. Between 5-6/1000 require specialized interventions and approximately 50 percent of these are patients during the first few months of life.1 A classification of various heart diseases based on severity is presented in the table below.

A declining trend in infant mortality rates is now being witnessed in almost every part of the world. This decline is almost entirely because of reductions in mortality from common childhood infections such as diarrhoea and pneumonia. This decline has allowed Congenital heart defects to surface as a significant health problem among infants and newborns in several parts of India and other low and middle income countries.2

The clinical spectrum of CHD varies from critical CHD that requires `immediate attention to minor CHD that needs occasional follow up. The broad categories of CHD along with the implications for survival and the appropriate treatment approaches are presented as Table 1 below.

Caring for CHD in the very young- Essential requirements

Care of children with heart defects is resource intensive and needs a high level of expertise among care providers and advanced equipment together with quality infrastructure and robust systems. It is also essential that the care providers work cohesively. The team is generally constituted by highly qualified health professionals that include pediatric heart surgeons, pediatric cardiologists, intensive care experts and, specially trained nurses. The most vulnerable group, newborns and infants, require the maximum resources and expertise.

Table 2 lists the requirements for development of comprehensive facilities for care of children with heart conditions. Figure shows essential components of pediatric heart care.

2. Why was Hridyam established?

In 2012, concerned that its IMR had been stagnant for so long, the Government of Kerala commissioned the Indian Academy of Pediatrics, Kerala Chapter to evaluate the causes of IMR in the state [6]. That study showed that infant deaths from infection and malnutrition had significantly declined, and that birth defects were the leading cause of infant mortality (30%). The study showed that a significant reduction in IMR would require treatment of birth defects. Among these, CHD represents the world's most common class of major birth defects, affecting one in 120 newborns. About one fourth of all CHDs are considered critical congenital heart disease (cCHD), which require a lifesaving procedure in the first year of life.

In 2012, the Government of India started the Rashtriya Bal SwasthyaKaryakram (RBSK), a national child health initiative for screening and treatment of childhood diseases and disabilities, including CHD. This program, administered by the National Health Mission (within the Ministry of Health and Family Welfare), provides funding and technical assistance to individual States. With the addition of funds and commitment by the Government of Kerala, adequate financial resources were available for an innovative population health approach to address the burden of CHD in the state.3

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Table 1: Broad Categories of Congenital Heart Disease Classified on Basis of Severity

Broad category	Implications for survival and treatment	Examples*
Critical CHD	Incompatible with survival without specific intervention in first few months of life	Transposition of the great arteries (TGA) , total anomalous pulmonary venous return (TAPVC), Pulmonary atresia associated with TOF or intact septum, critical PS etc.
Major CHD	Intervention is required, often in infancy, for optimal long-term outcome	Tetralogy of Fallot (TOF), large ventricular septal defect (VSD) and Patent ductus arteriosus (PDA), complete atrioventricular canal (AV canal), Anomalous left coronary artery from pulmonary artery (ALCAPA)
CHD that typically manifests at an older age and does not pose a mortality Risk	Diagnosis seldom made in early childhood; intervention required to prevent long-term problems in adulthood	Atrial septal defect (ASD), some forms of coarctation, less severe forms of aortic and pulmonary valve stenosis
Minor CHD	Long-term, symptom-free survival can be expected without any specific intervention in most cases	Small left-to-right shunts (ASD, VSD, PDA), bicuspid aortic valve

*These examples are not a comprehensive list of conditions; many conditions are not listed. Numerous combinations are possible.

Table 2: Essential elements for comprehensive pediatric heart care

No.	Checklist item
1	Quality Infrastructure & equipment
2	High level of skill among caregivers
3	Coherent teamwork
4	Supportive administration
5	Well-developed and mature referral base
6	Favorable economics and human development in the region
7	A system for charitable care
8	Sustainable systems and services: education and training, nursing
9	Constant focus on quality improvement
10	Ethical practice environment that is not totally profit driven

3. What is so unique about Hridyam?

a. First of a kind- Hridyam is perhaps the first example of a public health approach directed towards management of comprehensive congenital heart defects with a view to reduce its impact on infant mortality

b. Focus on infants and neonates: While there are examples of programs that provide for treatment of heart defects they are largely focussed on school going children and are therefore unlikely to have an impact on IMR. Hridyam is unique because it seeks to systematically identify and manage babies with critical CHD.

c. Attention to the care continuum- Grass roots approach: Hridyam seeks to strengthen health systems to enable to improve services at every stage of the care continuum of congenital heart disease from screening of heart defects at birth, stabilization and transport, provision of care including surgery or intervention and follow up care.

d. Transparency: The system allows data to be captured across every stage of the journey of a child with CHD. This can enable quality improvement by identifying key deficiencies

e. Robust IT platform: the web based platform created for Hridyam allows easy referral of babies with CHD, posting of expert opinions and scheduling of procedures across the state.

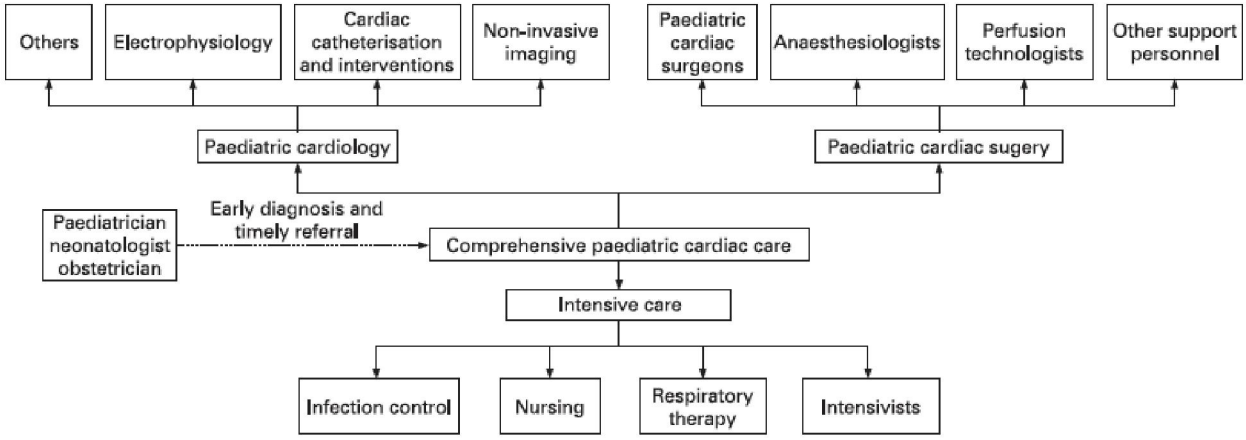

Fig.1. Components of a Comprehensive Pediatric Cardiac service

4.What has been accomplished through Hridyam?

a. State wide Network: An extensive state-wide network has been established that supports care of babies with CHD. This system seamlessly integrates primary care services with tertiary care centers that provide comprehensive pediatric care services. The DEIC managers and district coordinators have played a critical role here

b. CHD mortality reduction and IMR reduction:One of the main reasons for introducing the program was to reduce the infant mortality in the state to less than 10, i.e, single digit. This goal has been accomplished. The latest numbers as published in the Sample registration system bulletin of Govt of India reveals an IMR of 7 for the state of Kerala. 8 Figure 2 shows IMR trends for the state of Kerala over the last 3 decades

c. Education and Training of health workers: Over a 1000 Nurses have been trained for early detection. Additionally, several health workers and primary care providers have been sensitized towards the importance of early detection of CHD

d. Awareness: The awareness within the health sector and among the general public on CHD is unprecedented. This has allowed for very early diagnosis and timely referral of many babies with critical CHD.

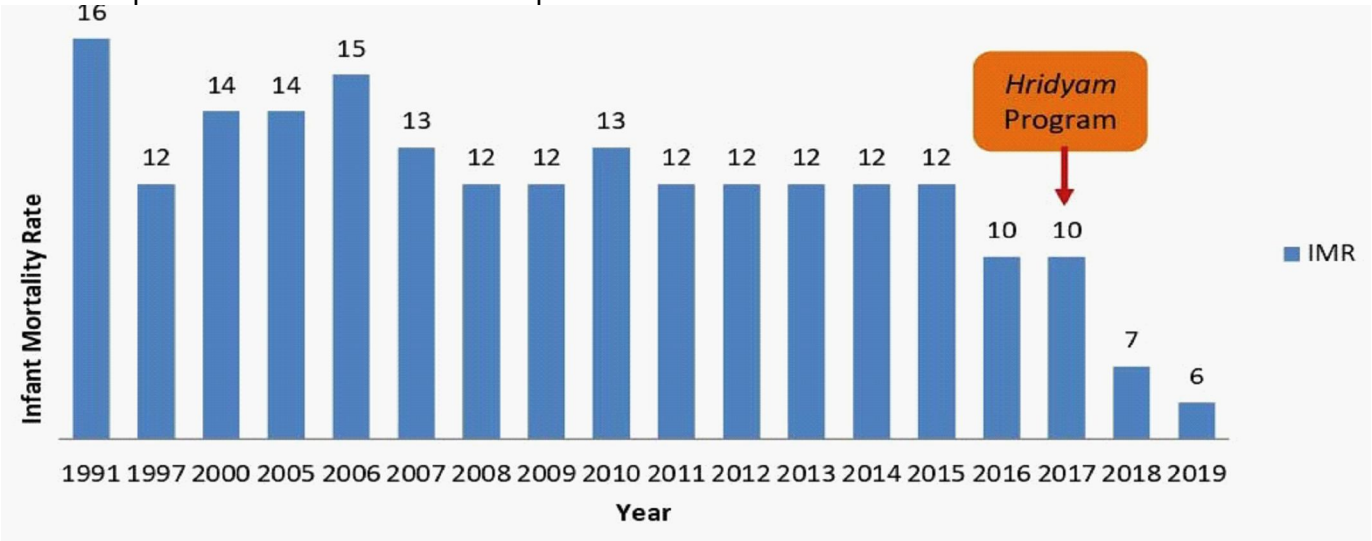

Fig.2. IMR trends in Kerala Since 19914

5. What has enabled the success of the program so far?

a. Exceptional leadership: The program was established and sustained in its initial stages because of exceptional leadership that paid close attention to all the early challenges and fixed all the bugs in the system

b. Dedication of DEIC managers: The team assembled for the implementation of Hridyam was singularly dedicated to the overarching goal of ensuring the best possible care for every baby with CHD. The DEIC managers , in particular, are exceptionally committed and much of the success of Hridyam can be attributed to them.

c. Commitment of all stake holders: Hridyam has unified all the care providers of CHD from both the government and private sector and motivated them to ensure the success of the mission.

d. Quality of primary care: The quality of primary health care in the state of Kerala was an essential ingredient for establishment and sustenance of Hridyam thus far.

6. What are the challenges that we face and how can they be addressed?

While the first five years of Hridyam has accomplished a great deal, perhaps, the greater challenge would be in ensuring its sustenance for the future. This would require attention to some important challenges that the program faces today:

a. Ensuring quality of care and building a robust system on accountability: It is vital to closely and comprehensively monitor every aspect of the program. This includes appropriateness of the indications for the procedures performed, effectiveness of screening, the quality of stabilization and transport, surgical and interventional outcomes. Without these checks in the system, there is a risk of the program losing its effectiveness.

b. Building capacity on early detection and preoperative stabilization in government hospitals: One of the main challenges is the fact that babies with CHD is the need for these babies to be taken care of as soon as the diagnosis is made. This requires training of pediatricians and neonatologists. The state medical colleges has established units that require to be trained for this purpose.

c. Need for continued training in prenatal and neonatal screening to ensure critical and correctable CHDs are not missed.

d. Prioritizing life threatening CHD in infants and neonates: Hridyam should not lose its core focus on saving young babies with critical CHD with a view to minimize IMR. While older children with CHD certainly need attention at some stage, it is not a public health priority. For example, if we chose to delay closure of a atrial septal defect by a few years, it should not significantly impact the future of the child. Examples of procedures that can wait include

   i. Atrial septal defects (ASDs)

   ii. Small VSDs that are referred for Device closures- These perhaps should not be covered by the scheme as their utility is controversial

   iii. Small patent ducts

   Scarce public health resources should not be diverted towards conditions that do not need to be prioritized. This will seriously drain the system. In most publicly funded programs (such as NHS) elective conditions are put on waiting lists and this is perfectly acceptable.

e. The cost of taking care of complex cases: Certain conditions should not perhaps be included for care under Hridyam because they can seriously burden the system. These are, fortunately, relatively rare. Below is a list of conditions that should not perhaps be included for care under Hridyam and this needs to be identified early and communicated at the DEIC level:

   - Pulmonary atresia with absent or diminutive pulmonary arteries and major collaterals
   - Single ventricle physiology with heterotaxy
   - Hypoplastic left heart syndrome
   - High risk Fontan palliation – more than mild AVVR, impaired ventricular function, elevated PVRI, small pulmonary arteries, heterotaxy
   - CHD with co-existent genetic syndromes or other organ system issues that are not

compatible with long term survival: Trisomy 13/18

- CHD with serious co-morbidities resulting in very high surgical risk (e.g. Untreated sepsis

f. Sustainability and long term health of partnering institutions: Partnering institutions (both government and private) have been stressed financially in implementing Hridyam. Their resources are finite and the compensation offered by Hridyam does not cover actual expenses. This challenge will eventually threaten the sustainability of the entire system. It is essential to be mindful of this challenge at all times and find ways to ensure that losses to participating hospitals are minimized.

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Case Report: Neglected Case of Developmental Dysplasia of Hip with Occult Contralateral Hip Dysplasia

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Abstract:

Background: Developmental Dysplasia of the Hip (DDH) encompasses a range of disorders that impact the proper development of the hip joint, typically diagnosed during infancy. However, overlooked occurrences in adolescents present distinct difficulties, particularly when accompanied by hidden contralateral hip dysplasia.

Case Presentation: We present a case involving a 12-year-old girl with an overlooked instance of bilateral DDH, exhibiting worsening gait issues and a discrepancy in limb length. Both clinical and radiographic assessments indicated significant dysplasia in the affected hip and undiscovered dysplasia in the contralateral hip, which was asymptomatic. A surgical procedure was planned to enhance hip stability and function while addressing bilateral hip issues.

Conclusion: This case emphasizes the significance of early identification and treatment of DDH to avert long-term complications. It also highlights the necessity for a comprehensive bilateral hip evaluation in cases of unilateral DDH to unveil hidden involvement and guarantee thorough management.

Introduction

Developmental Dysplasia of the Hip (DDH), also known as infantile hip dysplasia, is the primary cause of hip instability in children. The left hip is more frequently affected, accounting for 40 to 60% of cases, while bilateral involvement occurs in 12% to 20% of cases [5]. Although subtle dysplasia in the contralateral or otherwise normal hip can often be detected early through radiographic or ultrasound examinations, it can also become apparent during adolescence. It remains uncertain how common this occurrence is, whether acetabular dysplasia of the unaffected hip develops as the individual ages, or if it represents a very mild form of dysplasia that remains undetected until later in life. Limited research has clearly defined the changes that occur in the uninvolved hip with acetabular dysplasia. Understanding and raising awareness of this process is crucial for better prognosis, assessment, and management in the future.

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Case presentation

- A 12-year-old girl presents with complaints of progressive pain in the left hip and difficulty walking in the pediatrics OPD. The pain has been present for approximately 1 year and has been gradually worsening.
- The patient reports limping since early childhood, which was initially intermittent but has become persistent in the past few years. Parents recall delayed walking milestones, but did not seek medical attention earlier. Pain is localized to the left hip, worsens with physical activity, and is relieved by rest.
- No history of trauma or infections involving the hip joint.
- Past Medical History: No history of prior imaging or treatment for hip-related complaints.

Birth history: pre-term vaginal delivery, no known complications.

Physical Examination:

• Gait: Trendelenburg gait.

Right hip:

- Positive Galeazzi sign with limb length discrepancy.
- Limited range of motion, particularly in the abduction and internal rotation of the hip joint.
- Positive Trendelenburg test.
- No obvious swelling or redness.

Left hip:

Painful movement with no apparent deformity, but slightly positive anterior impingement test.

The patient was sent to the radiology department for a radiograph of the bilateral hip joint and an MRI of the pelvis.

On Pelvic X-ray (AP view): (Fig. 1)

- Right hip shows a high-riding femoral head with acetabular dysplasia.
- Shallow acetabulum with increased acetabular index angle.
- Left hip: femoral head epiphysis appears flattened and dysplastic.

An MRI of the pelvis was done.

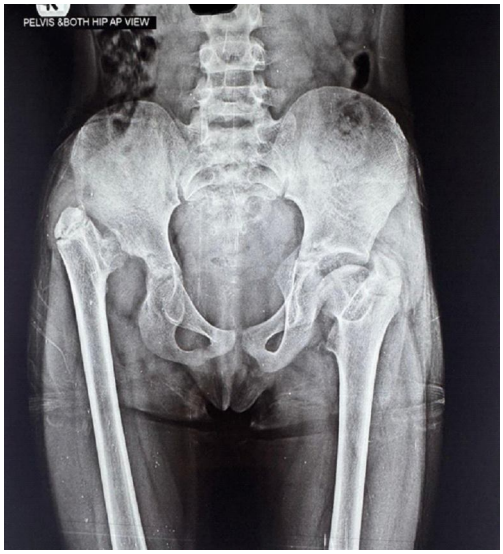


Fig 1. X-Ray of pelvis

For imaging and comparison of both the hips large FOV is required, for which a body coil is used; however, a dedicated hip coil was used to provide high-resolution images. Thin sections (3 or 4 mm) were used with minimal interslice gap. Standard imaging protocols in the hip included coronal T1W Spin echo, coronal short tau inversion recovery, axial T2 W Fast Spin Echo (FSE), fat-saturated oblique sagittal FSE, Proton Density (PD) FS, and axial FSE PD images. Additional protocols included axial three-dimensional spoiled gradient recall echo with fat suppression and post-contrast FS T1W.

Findings :

Right hip :

There is reduced bulk with fatty changes in right-sided gluteal muscles and iliopsoas muscle – Disuse atrophy (Fig. 7)

The right femoral head was displaced superiorly and laterally, with the femur head pointing anteriorly (femoral anteversion).

The right acetabulum appears shallow & dysplastic, and incongruent with the femur head. (Fig. 2 & 7)

Increased acetabular angle ~ 52 °. (Fig. 2)

Pulvinar fat hypertrophy is seen in its antero-inferior aspect. (Fig. 3)

The acetabular labrum appears thickened and interposed between the femur head and cartilage, causing its failure of reduction. (Fig. 4)

The iliopsoas tendon appears thickened and interposed between the femur head and acetabulum, failing reduction. (Fig. 5)

Left hip:

The left femoral epiphysis was flattened and widened, with mild widening of the acetabulum and surface irregularity (Fig. 6)

There was under-coverage of the femur head indicated by a central edge angle of ~34°. (Fig. 7) However, there was no migration of the femur head.

Discussion

Developmental dysplasia of the hip (DDH) necessitates careful assessment and treatment, Especially when it manifests in later childhood or adolescence. This case study features a 12-year-old girl with neglected right hip DDH and hidden dysplasia in the left hip emphasizing the challenges associated with treating unilateral DDH and the critical need for comprehensive screening of both hips.

Research indicates that it is not unusual for contralateral hip dysplasia to occur in individuals with unilateral DDH [9].Although generally mild, it frequently remains unnoticed during initial diagnoses or early childhood assessments, only becoming evident by skeletal maturity [5]. In this instance, while the patient exhibited significant acetabular dysplasia

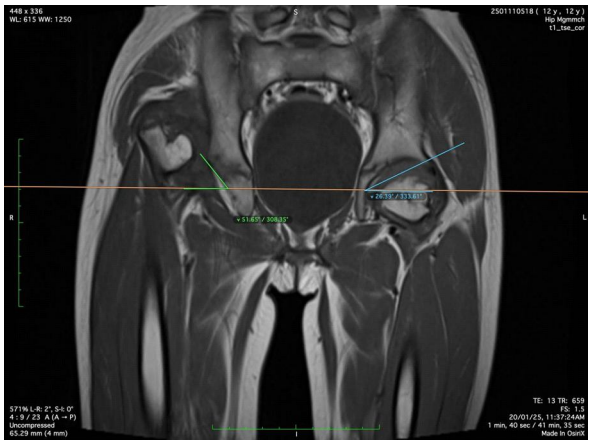


Fig 2. T1W coronal image showing normal acetabular index on the left side and abnormal on the right side. The acetabular index is the angle between the horizontal line through the sourcil and the line through the acetabular roof.[Right- 52 ° ; left 26 °] Normal range(range 18 ° to 32 °).
(Sourcil- area of subchondral condensation in the acetabular roof)

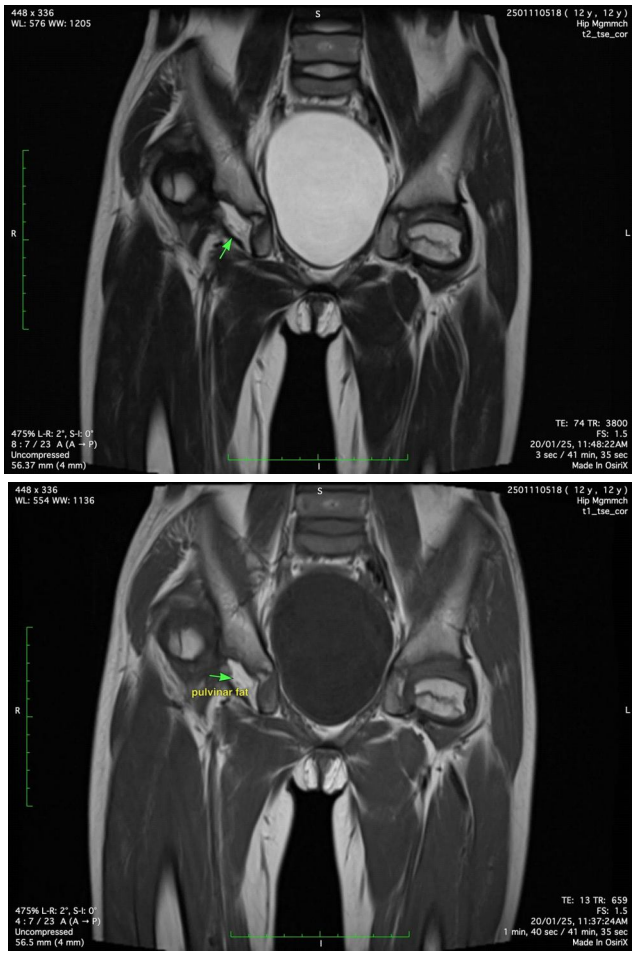


Fig 3a, 3b. Coronal T2 and T1-weighted images showing pulvinar fat hypertrophy in the right hip joint, cause of failed reduction.

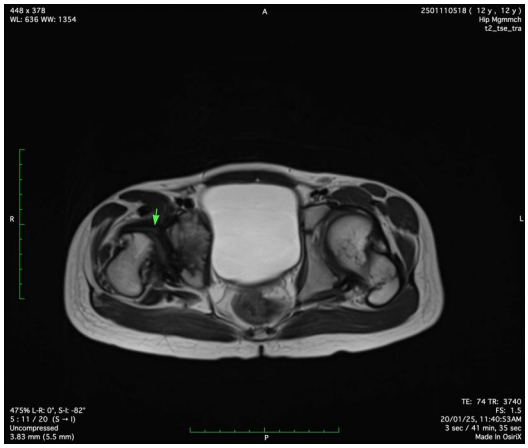


Fig 4. T2 axial image with right-sided DDH. The femoral head is displaced superiorly. The acetabular labrum is thickened and hypertrophied and slipped into the joint (arrow), which blocks the closed reduction.

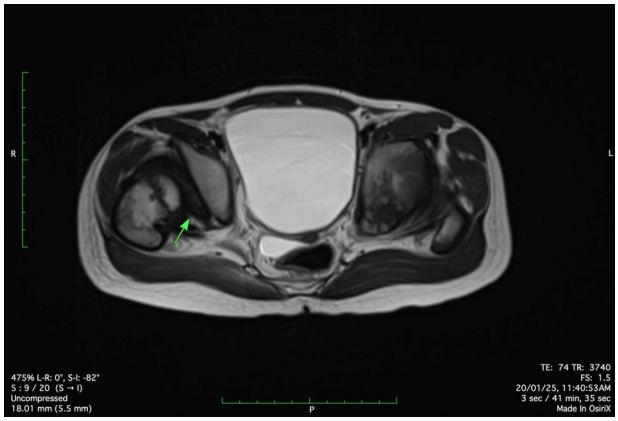


Fig 5. T2 W axial image depicting the interposition of the iliopsoas tendon on the right side (arrow), another cause of failed reduction

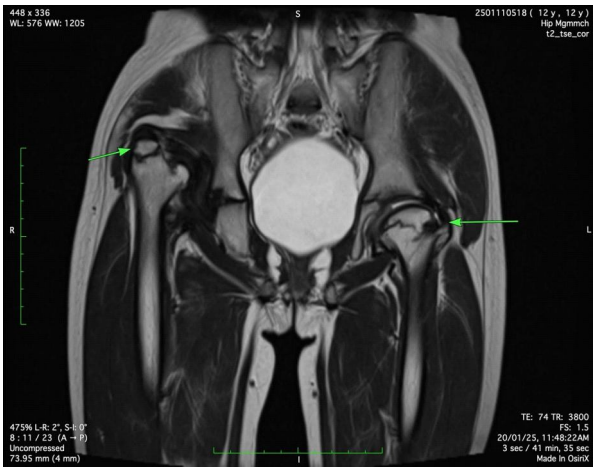
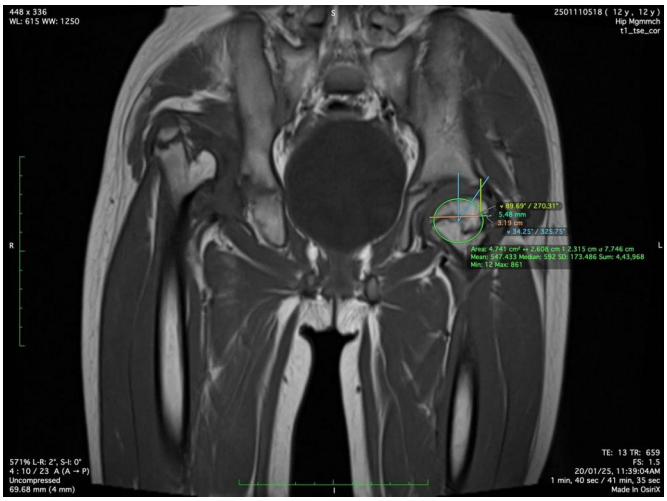


Fig 6. Right-sided DDH, T2 Coronal image: Head is dislocated Superiorly and laterally on the right side (arrow). The left femoral epiphysis is flattened and widened.



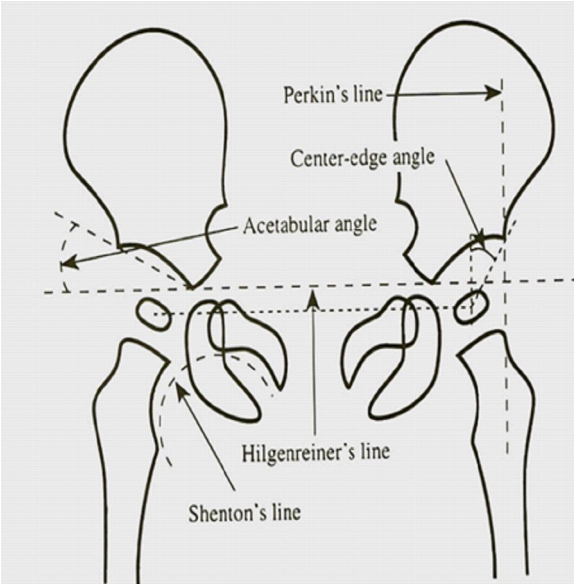


Fig 7. The center edge angle of Wiberg was measured as the arc between a vertical line through the center of the femoral head and a line drawn from the center of the femoral head to the outer edge of the acetabulum. It measures 32° (normally less than 20^0). The migration index of Reimers was measured as the percentage of the femoral head that was lateral to the lateral aspect of the acetabulum (16%) (normally less than 20%)

on the right side, an MRI of the left hip uncovered subtle abnormalities in the femoral epiphysis and acetabulum, indicative of occult dysplasia. Studies support the notion that mild dysplasia of the opposite hip in unilateral DDH might arise from irregular biomechanical forces, growth issues, or minor acetabular underdevelopment that was not recognizable during infancy.[4] These changes may result from altered mechanics due to extended compensation for the affected side, particularly in cases that have been overlooked. Detecting occult dysplasia necessitates comprehensive and precise imaging techniques. Standard radiographs, commonly used for DDH evaluation, may overlook minor acetabular dysplasia. Advanced imaging techniques like MRI offer a more accurate depiction of the acetabular and labral anatomy, as illustrated in this case. These observations highlight the necessity for close monitoring and imaging of both hips in patients with unilateral DDH, even if the other side appears unaffected during early evaluations. For those with late diagnoses or those unresponsive to treatment, MR imaging plays a vital role in the preoperative evaluation of femoral head deformities, acetabular dysplasia, and the extent to which the bony acetabulum covers the femoral head, along with the labral and musculotendinous structures surrounding the hip. Additionally, DDH may increase the risk of early osteoarthritis due to constant strain on the labrum coupled with cartilage injury.[3]

Conclusion

This case highlights the critical importance for early detection and treatment of DDH to prevent long-term morbidity. It also emphasizes the importance of evaluating both hips in patients presenting with unilateral symptoms, as occult dysplasia may exist contralaterally . A multidisciplinary approach incorporating vigilant monitoring and timely surgical intervention is of paramount importance in managing such complex cases to optimize patient outcomes.

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Merosin-deficient Congenital Muscular Dystrophyy: A Close Mimicker of Cerebral Palsy

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Abstract:

BACKGROUND - Congenital muscular dystrophies (CMD) are a heterogeneous group of disorders resulting in hypotonia, muscle weakness, and dystrophic or myopathic features on muscle biopsy. Merosin-deficient CMD is one of the variants occurring due to laminin a-2 gene (LAMA2) mutation which disrupt the normal basal lamina architecture and lead to muscle weakness in a limb-girdle distribution. CLINICAL DESCRIPTION- This case report is about a 14-year-old non-ambulatory boy with developmental delay, previously considered and treated as cerebral palsy because of a history of perinatal asphyxia. As the child grew up, features of myopathy developed, with raised creatine kinase (CK). MANAGEMENT AND OUTCOME- MRI brain showed classical changes of merosin-deficient CMD. Merosin staining of the muscle biopsy specimen was negative suggesting merosin deficiency. This case illustrates the severe end of the complete merosin deficiency phenotypic spectrum with greater language and cognitive delay as compared to usual CMD cases. CONCLUSION- Merosin deficient congenital muscular dystrophies are degenerative muscle diseases mainly inherited by autosomal recessive pattern.This variant has its simplicity in diagnosis by muscle biopsywhich aid in the diagnosis of the case outlined here as well.

Keywords : Congenital muscular dystrophy, Merosin deficiency, Laminin chain muscle biopsy.

Background :

Merosin deficient congenital muscular dystrophy (MDC1A) was the first congenital muscular dystrophy to be separately described with a definite genetic basis (mutation in LAMA2) [1,2]. The estimated relative frequency is between 10 to 30% of all CMD sub-types [3,4]. LAMA2 encodes the a-2 chains of the laminin-211 (also known as merosin), a major component of the basal lamina of Schwann cells and skeletal muscles [5]. Indeed, loss of function mutations of the LAMA2 gene results in muscular dystrophy, demyelinating neuropathy, and brain abnormalities. This results in Merosin deficient Congenital Muscular Dystrophy (MDC1A) also known as LAMA2-related muscular dystrophy (LAMA2-RD).It most commonly presents during infancy. Affected infants present with hypotonia, failure to thrive, respiratory difficulty, contractures and feeding problems. From 6 months of age, characteristic MRI changes could also be identified. Whilst cognitive function is usually normal, a small proportion of patients may have mental retardation or epilepsy [6]. MDC1A has been regarded as the most common congenital muscular dystrophy in Western countries, accounting for 30–40%of congenital muscular dystrophies (CMD)[7].

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Clinical Description:

14-year-old boy, born fifth in birth order, to a second degree consanguineously married parents, gravida5 para4 alive2 dead2 mother, came for evaluation of delay in achieving developmental milestones. He was delivered at full term by LSCS, birth weight of 2.8 kg and a history of perinatal asphyxia which required NICU care for 10 days. Parents noticed developmental delay as compared to the peers which became more apparent by the age of 2 years as evident by the inability to sit without support or speak not more than three four words. There was also a history of facial weakness developed from 6 years of age which caused difficulty in normal feeding. He had significant failure to thrive and was being diagnosed as cerebral palsy at local hospital, though no neuro-imaging was being done at that time. He had history of multiple hospital admissions for

recurrent pneumonia needing intravenous antibiotics. Recently at 14 years of age he presented to our hospital with pneumonia and for further neurological assessment. The child was severely wasted (fig.1) with weight 25 kg and length 135 cm (both were below the 3rd percentile). There was classical triangular facies with stiff tongue (fig.2,3). He had contractures at bilateral elbow, knee and wrist joints (fig.4). Both feet were fixed at plantar flexion (fig.5). Disuse atrophy of proximal muscles was evident on further examination, with hypotonia of all four limbs and trunk muscles as well. There was marked limb girdle weakness with power of 1/5 in lower limbs and 2/5 in both upper limbs with areflexia

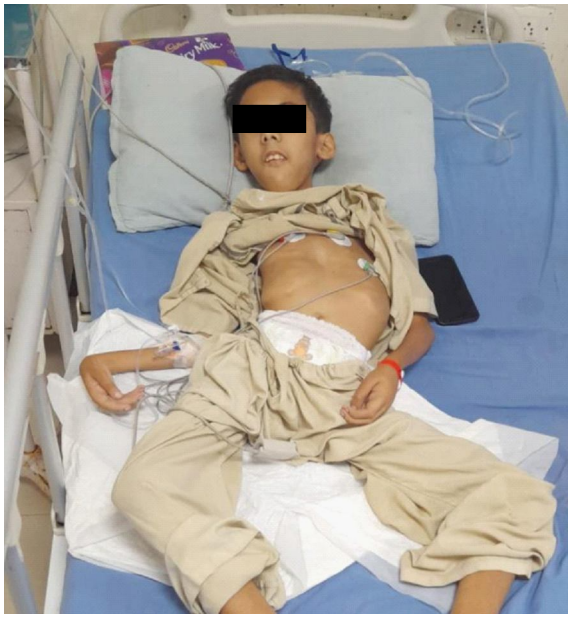

Fig 1: Severe wasting

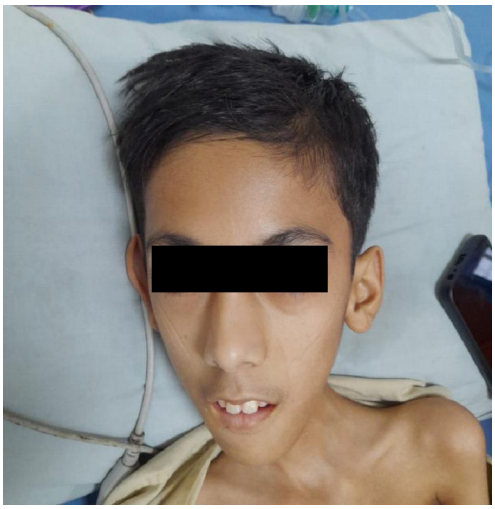

Fig 2: Classic myopathic facies

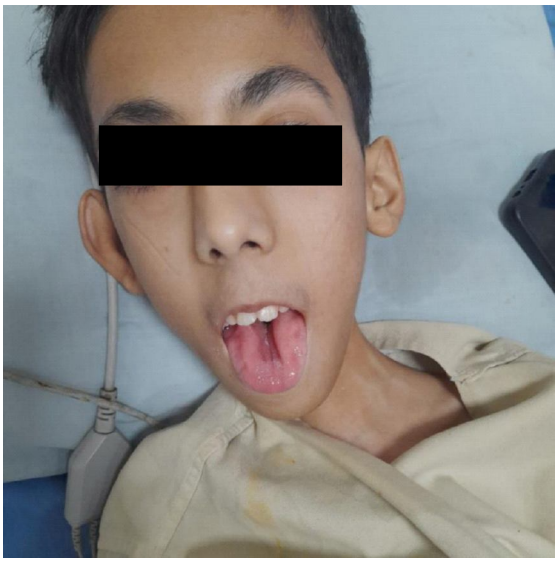

Fig 3: Stiff tongue

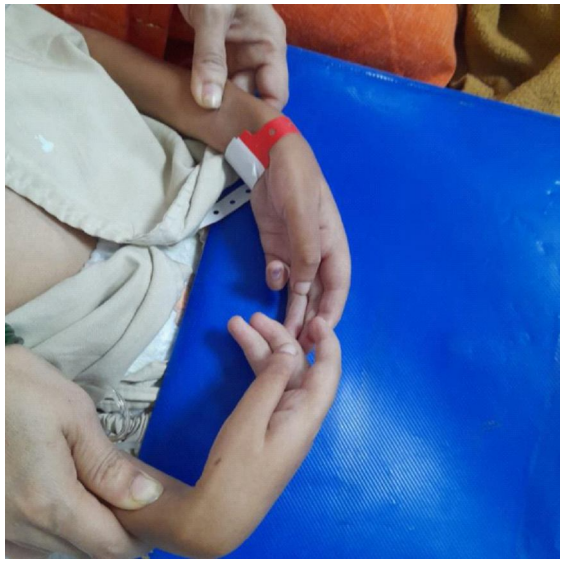

Fig 4: Wrist contractures

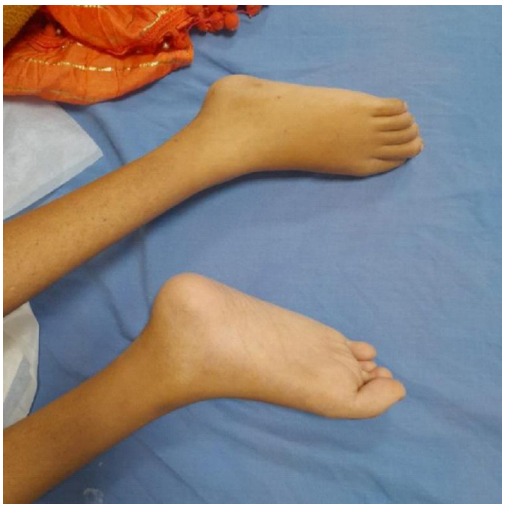

Fig 5: Plantar flexed foot

in all four limbs. Touch, proprioception and vibration sensation were intact.

Management And Outcome:

Chest X-ray revealed right sided pneumonia for which he required HFNC support for 3 days and a course of IV antibiotics, followed by oral, upon which resolution was seen. His serum creatine kinase levels were elevated up to 4000 U/L. A variety of metabolic investigations sent were normal. Cardiac workup was normal. Symmetric motor axonopathy and myopathy features were noted in needle EMG. MRI brain revealed T2 hyper-intensity and T1 hypo intensity involving the deep and periventricular white matter sparing the sub-cortical white matter in bilateral cerebral hemispheres along with pontine atrophy and mild cerebellar dysplasia. Biopsy of left quadriceps muscle done at an outside setup gave the impression of end stage histology with extensive replacement with hyalinized fibrous connective tissue. A muscle biopsy specimen stain with merosin showed no merosin uptake, indicating a complete loss of merosin protein. On the clinical background of the myopathic features, along with the classical MRI findings suggestive of congenital myopathy, and a merosin negative muscle staining, the diagnosis of merosin deficient CMD in a 14-year-old boy was reached upon. As of now, there is no definitive treatment for this variant. Supportive care in the form of physiotherapy and nutritional rehabilitation was initiated.

Discussion

Laminin are cross-shaped hetero-trimers constituted by the association of 3 different gene products, the a, ß and ? chains, and the most abundant structural non-collagenous glycoprotein in basement membrane. Laminin a-2 chain, which was originally called as merosin or laminin M-chain, has been identified as a tissue-specific laminin a-2 chain, and was first described by E. Engvall's group [8]. In human, the LAMA2 gene is located on 6q22–23 and consists of 64 exons [9]. Analysis of the laminin a-2 chain circular DNA or the LAMA2 gene itself showed that nucleotide substitutions, small deletions, or insertions induce complete merosin deficiency and a severe phenotype. The spectrum of the phenotypes of CMD patients with partial merosin deficiency is wide and is caused by a homozygous missense mutation, a missense mutation associated with a nonsense mutation, in-frame deletions in the LAMA2 gene, or nonsense mutations [10]. Laminina-2chain is rich in extra- synaptic, primary cleft, and junction fold of basement membrane [11]. It is assumed to play an essential role in basement membrane formation through multiple interactions with itself and other components [12]. It interacts with various cells through laminin receptors on cell membranes to control cellular activities such as adhesion, migration, differentiation, polarity, proliferation, apoptosis, and gene expression etc. Two merosin receptors are being identified, integrins and a non-integrins laminin receptor, called dystroglycan [13]. Both are thought to be critical to maintain normal muscle function. A study showed that reduction of a-dystroglycan (aDG) in myotubes in vitro results in reduced levels of laminin expression on their surfaces, an increase in cell death and TUNEL-positive nuclei [14].

Merosin-deficient CMD cases have been described as a relatively homogeneous group: severe neonatal hypotonia associated with joint contracture, inability to stand or walk, highly elevated serum creatine kinase (CK), alterations in somato-sensory and visual evoked potentials, cerebellar white matter involvement with abnormal signaling on brain imaging, but no abnormalities in cortex development or gyration are the features. Most patients have a normal intelligence and cognition. However, our patient had a greater language and cognitive delay than usual. In spite of the category of classical congenital muscle dystrophy, some children have been reported to show moderate mental retardation and epilepsy. Due to expiratory muscle weakness, secretion management is impaired because of ineffective expiratory flow during cough. Saliva and mucus may accumulate in the upper airways and favor local infections, which may then propagate to lower respiratory tract and the lungs leading to recurrent pneumonia in advanced stages. Usually, patients show no apparent myo-cardiopathy in spite of the lack of a-2 chain expression in heart. Laminin a-2 chain may be important for the selective filtration capability of the blood-brain barrier and the lack ofa-2 chain may cause impaired selective filtration, leading to leakage of plasma components and damage to the CNS [15].

Diagnosis of this condition has largely relied on immunocytochemical analysis of thea-2 chain in muscle biopsy specimens to confirm a loss of protein expression. Creatine kinase is often elevated in patients with congenital muscular dystrophy due to destabilization of the sarcolemmal membrane. Muscle biopsy shows dystrophic features: marked variation in fibre size with evidence of necrotic and regenerating processes, marked interstitial fibrosis, and adipose tissue proliferation. On electron microscopy, the basement membrane of muscle fibers is very poorly discernible and occasionally disrupted [16,17]. MRI brain shows striking white matter changes on T-2 weighted brain magnetic resonance imaging (MRI), which is diffuse, bilateral and quite symmetrical, appeared after the first 6 months of life, and persisted with time. Usually, this change is non- progressive. In addition to the white matter changes, some affected children show structural abnormalities, mainly involving the occipital cortex: occipital agyria, hypoplasia of cerebellum and pons, and ventricular dilation [18,19]. Brain MRI findings, which are found in almost all patients of merosin-deficiency, may be a valuable criterion for diagnosis of merosin deficiency in patients with classic congenital muscular dystrophy [20].

Muscular dystrophy is often viewed with diagnostic apathy as there is no curative treatment currently. Supportive care is essential in the form of physiotherapy to allow for maximal functional ability. Nutritional support is recommended due to severe wasting. Ventilatory support and tracheostomy maybe required in advanced stages. Management strategies also involve gene therapy but are mainly experimental as of now. Gene replacement is limited by the size of the LAMA2 gene (around 9 Kb), too large to be inserted in useful viral vectors. Gene editing, instead, has been successfully used to repair LAMA2 mutations [21]. Use of linker proteins i.e. mini-agrin and aLNNd is another emerging strategy. The first one was able to reconnect orphan laminin-211 receptors to the other laminin isoforms expressed in muscle and nerves and the second one to allow laminin polymerization [22] .The role of doxycycline might be linked to reduced cell death of immature Schwann cells and amelioration of Schwann cell differentiation [23]. Glatiramer acetate (GA), an agent for immune modulation has been shown to significantly improve mobility and muscle strength in the Lama2dy2J/dy2J mice and is under trial for human models now [24]. Prenatal diagnosis of the merosin-deficient congenital muscular dystrophy can be made by immune-cytochemical and molecular genetics studies of trophoblast [25].

Conclusion:

Merosin deficient CMD presents as hypotonia, respiratory and feeding difficulties in infantile age group and then progress to contractures, ambulatory difficulties and recurrent pneumonia in advanced stages. This case report outlines the importance of prompt and thorough workup for cases of developmental delay. Considering the greater language and cognitive delay than usual and history of perinatal asphyxia as in our patient, cerebral palsy might be a close mimic. Meticulous neurological examination and elevated CPK may point towards the myopathy domain and subsequent investigations leads to the diagnosis.

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LESSONS LEARNT:

1. Congenital muscular dystrophy has various subtypes.
2. Muscle biopsy with immuno-histochemistry and appropriate genetic study are required for the definitive diagnosis of the CMD subtype.
3. Management is mainly supportive.

DECLARATION OF PATIENT CONSENT:

The author has obtained consent from patient and parents regarding publishing of images and clinical information in this case report

FINANCIAL SUPPORT: Nil

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Registration Category	Currency	Early Bird (1st -30th April 2025)	Slab 1 (1st May-30th June 2025)
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