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Perinatal Society of Sri Lanka
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Towards healthier mothers and newborns

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Editorial

Navigating Sexual and Reproductive Health Amidst Global Economic Turmoil: A Sri Lankan Perspective

In an increasingly interconnected world, the ripple effects of global economic instability are being felt in nearly every domain of human development, including the often-overlooked sphere of sexual and reproductive health (SRH). The resulting constraints on national health systems are starkly visible as countries grapple with tariff wars, inflationary pressures, currency depreciation, geopolitical conflicts, and climate-related disasters. For Sri Lanka, a country already navigating a multifaceted economic crisis, the impact on SRH is profound and urgent.

Sexual and reproductive health is not a peripheral aspect of public health. It is foundational. It encompasses family planning, maternal and newborn care, prevention and treatment of sexually transmitted infections (STIs), adolescent health, and gender-based violence response services. These services are intrinsically linked to health equity, gender equality, and social development. Yet, in times of fiscal austerity and competing national priorities, SRH services are frequently among the first to face reductions in funding, accessibility, and political attention.

Globally, the consequences of economic instability on SRH are becoming increasingly visible. According to the United Nations Population Fund (UNFPA), supply chain disruptions have led to shortages of contraceptives and essential obstetric medications in numerous low- and middle-income countries. Simultaneously, donor fatigue and the reallocation of international development assistance have diminished global investments in SRH programs.

As a result, we are witnessing an increase in unplanned pregnancies, unsafe abortions, maternal mortality, and sexual violence—reversing gains made over decades.

Sri Lanka's situation signifies these broader trends yet is shaped by its unique context. The economic downturn since 2022 has significantly strained our health system. Public-sector health institutions, which serve the vast majority of Sri Lankans, are contending with severe shortages of essential supplies, including oxytocin, magnesium sulphate, antibiotics, and a range of contraceptives. Import restrictions, currency volatility, and constrained public expenditures have made procuring life-saving commodities increasingly tricky.

At the same time, human resources for health have also come under pressure. The migration of skilled health professionals, particularly nurses and midwives, has created service gaps in many peripheral areas, undermining maternal and newborn care continuity. Community-based services led by public health midwives, which have long been a strength of Sri Lanka's primary healthcare system, are increasingly overstretched and under-resourced.

Women in rural, estate, and underserved urban communities bear the brunt of this reality. Rising transport costs, limited clinic availability, and the inability to purchase necessities such as sanitary napkins or prenatal supplements contribute to worsening perinatal outcomes. Adolescent girls, in particular, are at greater risk of school dropout, early marriage, and unintended pregnancies, often with limited access to accurate information or supportive services.

Moreover, gender-based violence has risen amidst the socio-economic distress, while support services, shelters, counselling, and legal aid have not expanded in response. Social stigma and entrenched gender norms further complicate efforts to address these issues openly and effectively.

Despite these challenges, there are commendable examples of resilience and innovation. Digital health platforms are being piloted to provide SRH information and teleconsultations. Non-governmental organizations and professional associations have stepped in to fill service delivery gaps. Notably, the dedication of frontline workers, especially public health midwives and medical officers of health, has enabled some continuity in maternal and child health services under challenging circumstances.

However, these efforts must be supported and scaled. As healthcare professionals, researchers, and policymakers, we must advocate for a renewed commitment to SRH in our national recovery plans. Investments in SRH are not just ethical imperatives but also economically prudent. Each dollar spent on family planning and maternal health yields manifold returns in reduced healthcare costs, improved productivity, and enhanced quality of life.

To safeguard the gains made in perinatal and reproductive health, the following actions are essential:

1. **Ring-fencing SRH funding** within health budgets, even in times of fiscal constraint.
2. **Strengthening local manufacturing** and supply chains for essential SRH commodities to reduce import dependency.
3. **Supporting and retaining the health workforce**, especially in maternal and newborn care, through incentives and professional development.
4. **Expanding community outreach**, particularly to marginalized populations, via well-resourced primary healthcare networks.
5. **Integrating SRH into broader social protection and economic recovery policies**, recognizing its centrality to national development.

As Sri Lanka strives to rebuild its economy and strengthen its health system, sexual and reproductive health must not be an afterthought. It is a litmus test of our values, our priorities, and our commitment to human dignity. The lessons of this crisis should propel us towards building a more resilient, inclusive, and equitable healthcare system where every woman, adolescent, and family has the right and means to make informed choices and access essential care, regardless of economic circumstance.

Let this be the moment where we reaffirm our collective responsibility to protect not only life but the quality and dignity of that life through the lens of comprehensive sexual and reproductive health.

Dr. Surantha Perera
Managing Editor

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Presidential Address 2025

Enhancement of Quality of Perinatal Care through Maternal and Perinatal Death Surveillance and Response (MPDSR): A Vision for 2025 and Beyond

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A Turning Point for Maternal and Newborn Health

Sri Lanka's healthcare system has garnered global recognition for its impressive maternal and child health indicators. As a lower-middle-income country, it has outperformed many of its regional and economic peers, achieving near-universal institutional deliveries, high rates of antenatal care coverage, and an equitable public health network reaching across the island. Despite these achievements, Sri Lanka finds itself at a critical juncture where **further reductions in perinatal mortality** require not only continued effort but also strategic innovation and a bold transformation in the **quality of care** delivered to mothers and newborns.

In this context, the Perinatal Society of Sri Lanka (PSSL) has adopted the theme for 2025: "Enhancement of Quality of Perinatal Care through Maternal and Perinatal Death Surveillance and Response (MPDSR)." This theme encapsulates the Society's commitment to moving beyond quantitative targets and focusing on deeper systemic changes—particularly, the use of data and collaborative inquiry to shape clinical practice and health policy.

My address today outlines the rationale, strategic direction, and envisioned actions under this theme, serving both as a reflection of national priorities and as a call to collective responsibility among professionals and policymakers alike.

Historical Context and the Role of PSSL

The Perinatal Society of Sri Lanka was founded in 2001 by a group of passionate clinicians, led by Emeritus Professor Indrajee Amarasinghe. Beginning with just nineteen life members and eight ordinary members, the Society has since matured into a vibrant, multidisciplinary body recognized for its advocacy, technical leadership, and its integrative approach to perinatal health. The Society's foundation was built on the understanding that perinatal outcomes are influenced not solely by clinical care at birth but by a wide continuum of factors, from antenatal screening and management of maternal comorbidities to postpartum follow-up and neonatal care.

What distinguishes the PSSL is its inclusive membership, which now extends across obstetricians, pediatricians, neonatologists, community physicians, and also physicians involved in caring for maternal complications effecting the perinatal outcome. The membership is further strengthened by the involvement of nursing and midwifery group as the associate members. This multidisciplinary ethos has become the Society's trademark, promoting cross-sectoral problem-solving and innovation in service delivery. In recent years, the Society has also taken a leadership role in regional policy discussions, contributed to the development of national guidelines, and worked closely with the Ministry of Health and the Family Health Bureau to align clinical standards with emerging global benchmarks.

The PSSL has also forged strong working relationships with key development partners, including the World Health Organization (WHO), UNICEF, UNFPA, and the World Bank, whose support has been instrumental in catalyzing quality improvement initiatives, expanding training infrastructure, and facilitating research into maternal and newborn health.

Global and National Burden of Perinatal Mortality

Perinatal mortality, defined by the WHO as the combination of stillbirths (from 28 weeks of gestation) and early neonatal deaths (within the first 7 days of life), remains one of the most persistent challenges in global public health. Each year, approximately **2.4 million newborns** die, and an additional **2 million stillbirths** occur. Among these stillbirths, **over 40% take place during labor**, known as intrapartum stillbirths. These deaths are particularly tragic, as they are often preventable with timely and skilled obstetric care. The burden of perinatal mortality is unevenly distributed: low- and middle-income countries bear over 98% of these losses, with Sub-Saharan Africa and South Asia accounting for the majority.

In Sri Lanka, official data reveals a different, more nuanced picture. The **maternal mortality ratio (MMR)** stands at an impressive **25 per 100,000 live births**, among the lowest in South Asia. The success is attributed to near-universal institutional births, skilled birth attendance, and a well-organized maternal health service delivery structure. However, **neonatal mortality** has not demonstrated a proportional decline. Over the last decade, rates have remained relatively stagnant, hovering around **5 to 6 deaths per 1,000 live births**, with a significant portion occurring within the first 48 hours post-delivery.

This stagnation signals that basic coverage and access alone are no longer sufficient. Instead, **quality of care**—both in terms of clinical practice and health system responsiveness—must now take center stage. Moreover, disparities remain: women from remote, estate, or conflict-affected regions continue to experience higher rates of adverse outcomes, partly due to delays in accessing emergency services and inconsistencies in clinical decision-making.

The Sustainable Development Goals and Sri Lanka's Commitments

The global health community, through the **Sustainable Development Goals (SDGs)**, has established ambitious yet achievable targets for maternal and newborn health. **SDG 3.1** calls for the reduction of the global maternal mortality ratio to less than 70 per 100,000 live births by 2030, with Sri Lanka targeting an even more ambitious **MMR of 16 per 100,000 live births**. **SDG 3.2** focuses on ending preventable deaths of newborns and children under five years of age, with neonatal mortality to be reduced to **at least 12 per 1,000 live births, globally**.

In addition, **SDG 3.8**—often overlooked—emphasizes universal health coverage, not only in terms of access but also in terms of **quality, safety, and affordability**. While Sri Lanka has made commendable strides in reaching the first two dimensions, further effort is required in ensuring that quality benchmarks are met across all sectors and all geographies.
PSSL's 2025 theme directly supports these global and national commitments, particularly by promoting systems and strategies that generate actionable insights and catalyze reform where it is most needed.

Maternal and Perinatal Death Surveillance and Response (MPDSR): The Engine for Quality Improvement

One of the most transformative tools available for addressing perinatal mortality is the **MPDSR system**, a WHO-endorsed initiative aimed at identifying, reviewing,

and responding to maternal and perinatal deaths. Unlike passive data reporting systems, MPDSR is a **cyclical, continuous process** that emphasizes learning, accountability, and preventive action.

The system begins with the notification of a death, which triggers a multidisciplinary review at the institutional or district level. These reviews examine the medical, social, logistical, and systemic factors that may have contributed to the outcome. Rather than assigning blame, the process fosters a culture of openness and shared responsibility. Crucially, each review concludes with a set of specific recommendations—ranging from changes in clinical protocol to improvements in referral systems or equipment availability—that are tracked and evaluated over time.

Sri Lanka has made strides in institutionalizing MPDSR, with numerous hospitals and districts now conducting regular reviews. However, gaps remain in coverage, consistency, and follow-up. In 2025, PSSL aims to expand MPDSR to **all institutions providing maternal and neonatal care**, improve the quality of reviews through training and mentorship, and establish national coordination mechanisms that can translate findings into policy and practice.
International experience demonstrates that effective MPDSR implementation can reduce perinatal mortality by 20–30%, especially when combined with community engagement, health worker empowerment, and political commitment.

Strategic Priorities for 2025

The Perinatal Society's vision for 2025 is not merely aspirational—it is grounded in a detailed roadmap that encompasses systemic, clinical, and educational components. The Society is working towards:

1. **Institutionalizing MPDSR Across All Levels of the Health System**

   This involves advocacy with hospital administrators, training programs for clinical staff, and strengthening health information systems for timely death notification and data flow.

2. **Extending Confidential Enquiries into Maternal Deaths (CEMD)**

   Unlike MPDSR, which is primarily responsive, CEMD adopts a forensic, research-oriented approach to understanding patterns and systemic issues. PSSL aims to support the Ministry of Health in developing a nationwide CEMD platform, modeled on successful examples from the UK and South Africa.

3. **Building Public-Private Partnerships for Quality Assurance**

   The private healthcare sector accounts for a significant portion of deliveries in Sri Lanka. PSSL is engaging with private institutions to harmonize protocols, share data, and ensure that all women receive consistent, evidence-based care regardless of where they seek services.

4. **Promoting Research and Data Use in Policy Making**

   The Society will commission and support research into topics such as maternal mental health, congenital anomalies, and low birth weight—areas that are underrepresented in existing data sets.

5. **Investing in Education and Professional Development**

   From webinars to conferences, mentorship to simulation training, the PSSL will expand its offerings to ensure that every provider of perinatal care is equipped with the latest knowledge and tools.

Conclusion: A Call to Collective Responsibility

The journey toward reducing perinatal mortality and improving maternal health is no longer one of expanding access—it is a journey of **refining quality, eliminating disparities, and building resilient systems**. The PSSL believes that every maternal or neonatal death is not just a tragedy—it is a message. A message that compels us to look deeper, act faster, and care better.

As we embark on the year 2025, the Society invites all stakeholders—clinicians, administrators, policy makers, community leaders, and development partners—to join hands in this mission. Let us make Sri Lanka not only a success story in numbers, but a beacon of **equity, compassion, and clinical excellence** in perinatal care.

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Current Practice

Newborn screening in Sri Lanka

M.N. Lucas[1,2]

Background

Newborn screening (NBS) aims at identifying conditions that can affect a child's long-term health and survival before babies become symptomatic (1, 2). The foundation for newborn screening was laid by Dr Robert Guthrie in 1960, with the discovery of the dried blood spot to diagnose phenylketonuria (3).

NBS was initiated in Sri Lanka in 2008 with the introduction of screening for congenital hypothyroidism regionally. Island wide newborn screening for congenital hypothyroidism was initiated in 2016 (2). This was followed by the introduction of pulse oximetry screening in 2017 and screening for congenital deafness in 2019 (2,4). In 2022, WHO released a guideline on recommendations for universal newborn screening for eye abnormalities, hearing impairment and neonatal hyperbilirubinaemia (5). A pilot project to streamline newborn screening was conducted by the Ministry of Health in collaboration with the WHO in 2024 in 5 hospitals namely, De Soysa Hospital for Women, Colombo, Castle Street hospital for Women, Colombo, German Friendship hospital, Galle, Teaching Hospital Kurunegala and Teaching Hospital Jaffna. This pilot project included introduction of screening for eye abnormalities, expanding pulse oximetry screening for critical congenital heart disease to include screening for all congenital heart diseases and revamping screening for congenital hearing impairment.

Newborn screening comprises of six essential components.

1. Education - education of parents, health professionals and public
2. Conducting newborn screening - parental counselling, time sensitive screening, testing, and reporting
3. Early follow up – tracking, referral for diagnostic testing
4. Diagnosis – confirmatory testing, counselling parents, referral for early intervention
5. Management – commencing early intervention, counselling, follow up.
6. Evaluation – outcome monitoring and quality assurance

While provision of education, pretest counselling, conducting newborn screening, posttest counselling and referral for diagnostic testing are conducted by the hospital where the baby was born (delivery hospital), diagnostic testing, commencement of early intervention and follow up are conducted by the nearest referral hospital. Both delivery and referral hospitals should have means of monitoring the outcomes of newborn screening to ensure quality assurance (6).

Healthcare staff responsible for antenatal education at the delivery hospital incorporate information about newborn screening including its importance, timing, procedure as well as interpreting its outcome into the existing antenatal health education sessions to increase awareness among prospective parents.

Pretest counselling is conducted in the postnatal ward, either as a group or

individually after the baby's birth, prior to conducting newborn screening. Newborn screening is done after the completion of 24 hours of age, (or nearest to 24 hours) at the delivery hospital, prior to the discharge of the newborn from the delivery hospital, while the baby is calm, preferably after a feed, at the bed side, after ensuring hand hygiene by the health care staff. Posttest counselling is done individually with regard to the test outcomes.

Newborn screening in Sri Lanka: the current context

At present, newborn screening in Sri Lanka is conducted via newborn examination in addition to pulse oximetry for congenital heart disease, serum TSH for congenital hypothyroidism and otoacoustic emission (OAE) test for congenital hearing impairment.

Newborn examination

Complete examination of all newborns within 24 hours of age is a national standard for newborn care (7). Neonatal examination includes screening for multiple pathological conditions as shown in table 1.

Table 1: Newborn screening embedded within the neonatal examination

Physical signs	**Interpretation**	**Intervention**
Pallor (palms and soles)	Anemia	Look for bleeding / other causes / start iron supplements
Yellow discoloration in skin	Neonatal jaundice	Look for cause / commence phototherapy based on serum bilirubin levels
Increased respiratory rate and / or recessions and /or grunting and /or nasal flaring	Respiratory distress requiring treatment	Look for cause / commence respiratory support
Peripheral cyanosis	Hypothermia (commonly)	Measure temperature / rewarm the baby / look for other causes
Heart murmurs	Congenital heart disease	Isolated murmurs without risk factors, without any other clinical abnormality in the cardiovascular system with normal pulse oximetry should have a Paediatric cardiology referral if the murmur persists beyond 6 weeks of age Presence of risk factors / any other clinical abnormality in the cardiovascular system or abnormal pulse oximetry indicates a Paediatric cardiology referral prior to discharge
Absent / reduced femoral pulses	Coarctation of aorta	Paediatric cardiology referral
Central cyanosis without respiratory distress	Congenital cyanotic heart disease	Paediatric cardiology referral
Central cyanosis with respiratory distress	Lung pathology	Look for cause / commence respiratory support

Table 1: Newborn screening embedded within the neonatal examination continued.

Physical signs	**Interpretation**	**Intervention**
Hepatomegaly, splenomegaly	Congenital infection / hemolytic disease of the newborn / storage disease / malignancies	Ultrasound scan abdomen / Look for cause
Ballotable kidneys	Renal disease – polycystic kidney disease / multi-cystic / dysplastic kidneys / hydronephrosis	Ultrasound scan abdomen and paediatric nephrology referral
Undescended testis	Undescended testis	Paediatric surgical referral
Abnormal placement of the urethra	Hypospadias / epispadias	Paediatric surgical referral
Virilization of genitalia	Congenital adrenal hyperplasia (CAH)	Check capillary blood sugar, serum electrolytes - if positive do serum 17 OHP and USS abdomen to visualize ovaries / uterus and commence treatment if results are suggestive of CAH
Hypertonia / hypotonia with exaggerated or diminished deep tendon reflexes	Neuromuscular disease	Paediatric neurology referral
Lumps, hemangiomas, tuft of hair, over the spine	Spina bifida	USS spine and paediatric neurology referral
Inability to abduct hip completely	Congenital dislocation of the hip	Urgent USS hip and orthopedic referral
Inability to see the depth of the sacral dimple	Sacral sinus	USS spine and paediatric neurology referral
Absent anus	Imperforate anus	Paediatric surgical referral
Suture separation with large fontanelles	Macrocephaly, commonly hydrocephalus	USS brain followed by a paediatric neurology referral
Overriding sutures with very small fontanelles	Craniosynostosis	Paediatric neurology referral
Changes in occipitofrontal circumference (OFC) disproportionate to length	Macrocephaly (increasing OFC) Microcephaly (decreasing OFC)	Paediatric neurology referral
Absent / diminished red reflex	Cataract / retinoblastoma	Ophthalmology referral
Deficiencies in the lip and the palate	Cleft lip and palate	Orthodontic / plastic surgery referral
Dysmorphic features: flat nasal bridge / simian crease / sandal gap, thick nuchal fold, hypotonia	Down syndrome	Karyotype and Genetic referral Start physiotherapy Paediatric cardiology referral ENT referral Ophthalmology referral

Table 1: Newborn screening embedded within the neonatal examination continued.

Physical signs	Interpretation	Intervention
Dysmorphic features: prominent occiput, micrognathia, rocker bottom feet, overlapping fingers with a fisted hand	Edward syndrome	Karyotype and Genetic referral Counsel parents regarding poor prognosis
Neck lump in midline	Goitre / thyroglossal cyst	Paediatric surgical referral
Neck lump on the side	Sternomastoid tumours / branchial cysts / cystic hygroma	Paediatric surgical referral
Asymmetrical Moro reflex	Humerus fracture / clavicle fracture / Erb's palsy	Orthopedic referral for fractures Rheumatology referral for rehabilitation for Erb's palsy

Newborn Screening for congenital heart disease (CHD)

CHD are the commonest of all birth defects and account for up to half the deaths among congenital malformations. CHD have a prevalence of about 9 per 1000 live births, where 2 per 1000 are life threatening and ends up with circulatory collapse and death. These life-threatening CHD are known as critical congenital heart disease (CCHD) and are defined as cardiac lesions that require surgery or cardiac catheterization within the first year of life to prevent death or severe organ damage (8-12). A Sri Lankan study revealed that cardiac murmurs were heard in 0.6 – 8.6% of otherwise well looking newborns, where 37% were diagnosed with CHD and 2.5% with CCHD (12). A thorough clinical examination, as well as prenatal USS, can detect only 50-60% of CHD. The addition of pulse oximetry to a thorough clinical examination increases the detection rate to 92% (13).

While pulse oximetry is already being used to detect CCHD, the updated newborn screening guideline aims at detecting all CHD and recommends the addition of the ALT (ask, look and test) approach, where all newborns are screened for risk factors (see table 2) (**A**sk) and examined for clinical features (**L**ook) (see table 3) that increase the risk of CHD, in addition to being assessed with pulse oximetry (**T**est).

Table 2: Risk factors identified in the history *(adapted from Ford et al 2022) (14)*

Risk factors	Cardiology referral
Prenatal ultrasound suggestive of congenital heart disease	Before discharge – irrespective of presence of clinical features or pulse oximetry findings
Family history of congenital heart disease or hypertrophic cardiomyopathy Family history of inborn errors / lysosomal storage diseases Maternal drug history: Lithium, valproic acid, other antiepileptics, mycophenolate mofetil, warfarin, ACE inhibitors, retinoic acid, systemic vitamin A	Before discharge - only in the presence of clinical features or abnormal pulse oximetry After discharge – in the absence clinical features and normal pulse oximetry

Maternal diabetes in the first trimester Maternal infections in the first trimester Maternal obesity	Indicated only in the presence of clinical features of CHD or abnormal pulse oximetry

Presence of risk factors indicate cardiology referral in the presence of abnormal clinical features or abnormal pulse oximetry.

Prenatal fetal echo suggestive of CHD is an exception, that needs echocardiography before discharge irrespective of the findings in clinical features or pulse oximetry.

Maternal diabetes, maternal infection and maternal obesity indicate cardiology referral only if the baby has abnormal clinical features or abnormal pulse oximetry.

Table 3: Clinical features suggestive of CHD and urgency of cardiology referral *(adapted from Ford et al 2022) (14)*

Clinical feature	**Cardiology referral Timing, Urgency, and Indication**
Dysrhythmias Central cyanosis /recurrent peripheral cyanosis that does not improve with oxygen Sternal / anterior chest wall anomalies	**Before discharge** Risk factors present (table 1) / Positive pulse oximetry screen (Fig 8)
Abnormal heart sounds - • S2 – loud / soft / split • S3 • S4 • Clicks	**Within one week after discharge** No risk factors (table 1) and Negative pulse oximetry screen (Fig 8) Or **Before discharge** Positive for risk factors (table 1) / Positive pulse oximetry screen (Fig 8)
Pathological murmurs • Widespread murmur all over precordium • Long murmurs – pansystolic • Harsh murmurs • Intensity Grade 3 or more • Thrills • Diastolic murmurs	**Before discharge** No other clinical abnormalities and No risk factors (table 1) and Negative pulse oximetry screen (Fig 8)

Screening for risk factors and detection of abnormal clinical features is done by the medical officer at the time of the neonatal examination.

All abnormal clinical features indicate a cardiology referral. Management of neonates with a murmur is given in figure 1.

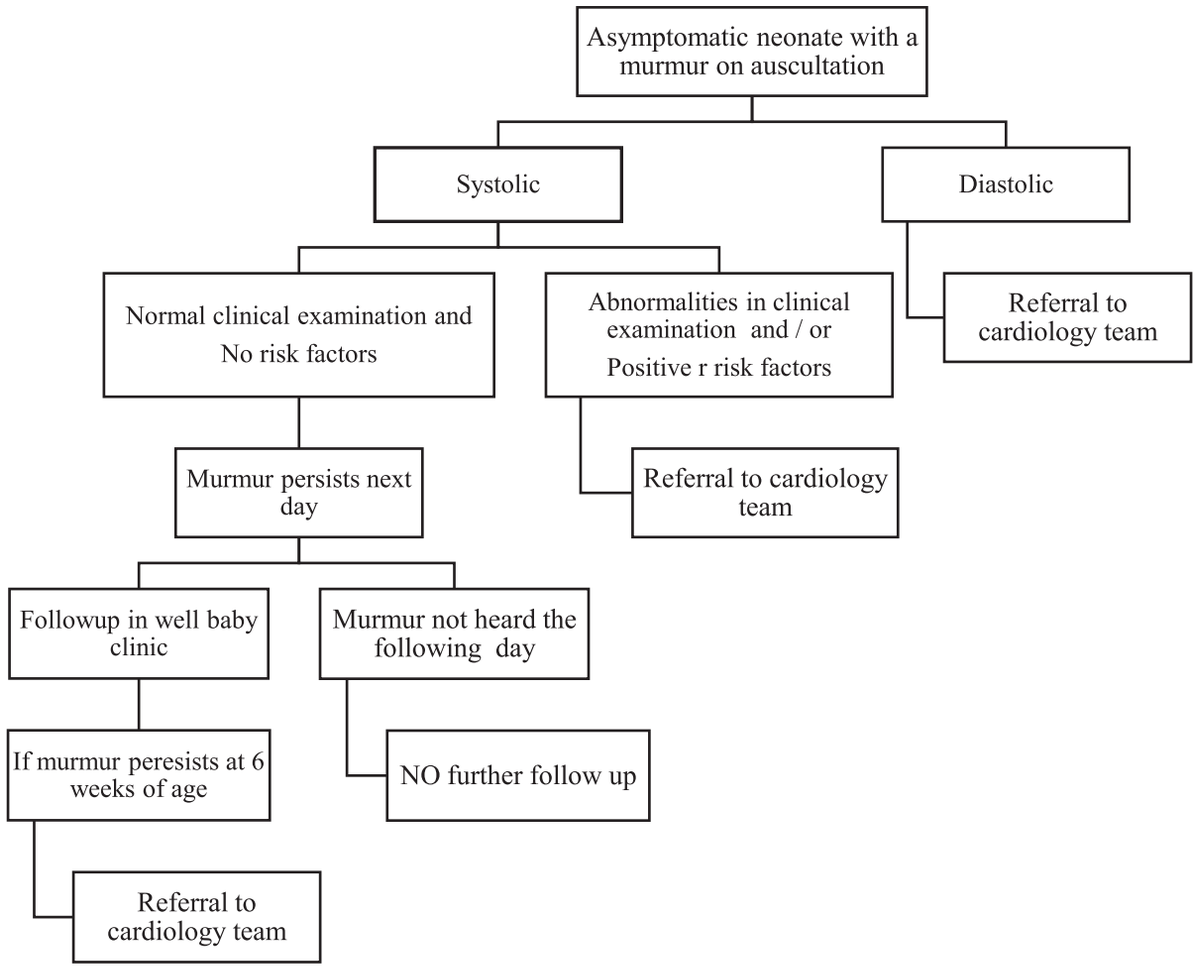

Figure 1: Indications to do a cardiology referral in a newborn with a murmur *(adapted from Ford et al 2022) (14)*

Pulse oximetry is conducted at the newborn's bedside, by a nursing officer preferably after completion of 24 hours, as this minimizes false positives that are commoner within the first 24 hours of life. Screen positives obtained before 24 hours of age, should be repeated after 24 hours of age, prior to cardiology referral if the baby is stable. This should be done only in babies on room air. Bright lights including phototherapy lights should be turned off to minimize interference. Limb should be clean, dry, free of motion, and well perfused prior to taking the measurement. The sensor and the detector should be opposite each other and should be wrapped tightly to minimize light exposure from outside. Do not apply plaster or hold the probe while taking the measurement. Management according to the pulse oximetry is given in figure 2.

Pulse oximetry is measured in the right hand as well as the foot. Right hand is selected as it receives blood supply before the aorta is connected to the ductus arteriosus, enabling the measurement of preductal saturation, compared to the other 3 limbs that receive their blood supply after the ductus connects to the aorta. Therefore, preductal saturation is measured using the right upper limb, whereas post ductal saturation is measured using the lower limbs. Difference between the pre and post ductal saturation is expected to be ≤3% in the absence of a duct dependent circulation in a baby with cyanotic heart disease. Presence of a duct dependent circulation will create a difference between

the pre and post ductal saturation >3%, where the mixing of deoxygenated blood entering via the ductus arteriosus will lower the post ductal saturation while maintaining the preductal saturation, thereby increasing the pre and post ductal difference to >3%. Overall saturation is expected to be ≥95%. While one reading of saturation <90% is taken as a positive screen, saturation readings between 90-94% or > 3% difference in pre and post ductal saturation, need confirmation with two repeat measurements one hour apart, i.e., 3 readings taken one hour apart, to be considered as a positive screen. Pre and post ductal saturation difference > 3% after 3 readings, each taken one hour apart warrant a cardiology referral. However, if the baby deteriorates awaiting cardiology assessment, a prostaglandin e2 infusion should be started to maintain the patency of the ductus arteriosus until the cardiology assessment. Management following the measurement of pulse oximetry is depicted in figure 2.

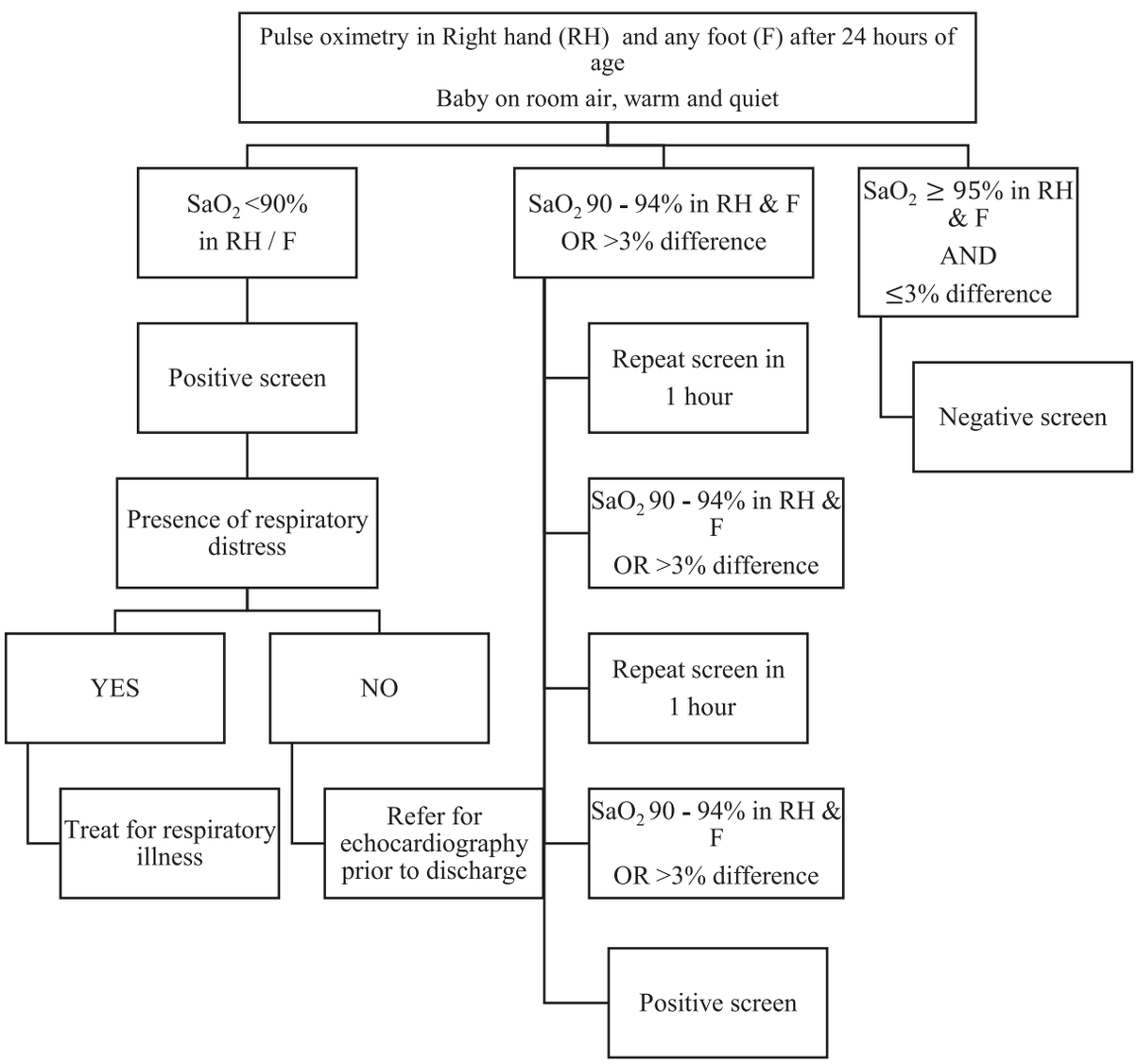

Figure 2: Management according to pulse oximetry findings *(adapted from the Ministry of Health circular 19.04.2017 on 'Newborn screening to detect CCHD) (15)*

Newborn Screening for congenital hypothyroidism

Untreated congenital hypothyroidism is the most common cause for intellectual disability. Early detection and treatment virtually eliminate the risk of intellectual disability (16). At present all newborns are screened prior to discharge. In 2021, incidence of congenital hypothyroidism was found to be 1 in 1289 following the screening of 239,737 newborns with 84% screening coverage. Blood is collected on to the blood spot marked on the filter paper, from the newborn via heel prick. Puncture site is disinfected using 70% alcohol and allowed to air dry. Warming the foot will increase the blood flow. The lateral aspect of the heel is punctured not exceeding 2mm by a sterile lancet / heel incision device. The first blood drop is wiped away with sterile cotton / gauze, while applying gentle pressure to form the second blood drop, that is directed towards the printed circle marked on the filter paper. A large blood drop needs to form and fall on the filter paper, so that it fully soaks and completely fills the printed circle. Excessive milking the puncture site is not recommended as it increases the risk of hemolysis and mixture of tissue fluids. The filter paper should not be pressed against the puncture site prior to the blood drop falling on the filter paper, as this would amount to an inadequate amount of blood to stain the filter paper on both sides. Blood should only be applied to one side of the filter paper. The filter paper is air dried and sent to the hospital laboratory that will assess the sample quality and then send the samples to the central laboratory (Medical Research Institute and Nuclear Medicine Unit, Karapitiya) within 3 days of sample collection. If the hospital laboratory does not send the sample to the central laboratory on the same day, samples should be refrigerated at 8^0C until it is transported, as moisture is detrimental the stability of the specimen.

The central laboratory quantifies serum TSH in the dried blood spot and communicates the test results to the MOH via e mail / telephone who in turn will inform the parents immediately and refer them to the nearest hospital with a paediatrician who should arrange confirmatory testing and initiate treatment with thyroxine (6). If the capillary TSH is ≥ 40 mIU/L, treatment should be commenced immediately after sending a confirmatory sample for freeT4(fT4) and TSH. If the capillary TSH is 20-40 mIU/L, treatment can be withheld until the next day, provided the fT4 and TSH reports are available the next day. If unsure when reports will be available, start treatment immediately after taking sample for fT4 and TSH. If the venous fT4 is below the reference range and or the TSH > 20 mIU/L treatment should be started immediately. Treatment is commenced with levothyroxine (L-T4) 10-15 micrograms/day. If, TSH is between 6-20 mIU/L with an age appropriate fT4 in a well-baby, repeat fT4 and TSH in 2 weeks (17).

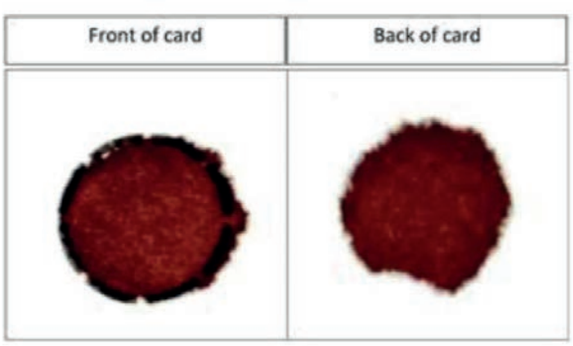

Figure 3: *Satisfactory dried blood spot on front and back of the filter paper (adapted from the Guidebook for healthcare staff. National newborn screening programme for congenital hypothyroidism 2023)(6)*

Newborn screening for congenital hearing impairment

Congenital hearing impairment occurs in every 3 to 5 per 1000 live births with severe to profound hearing impairment in every 1 per 1000 live births (18,19). Hearing impairment is commonly sensorineural, and results in irreversible deficits in communication, psychosocial skills, literacy leading to academic underachievement, unemployment, and psychological distress (19, 20). It is detected by parents / caregivers commonly around 2 years of age, typically due to delayed language development. The critical time for the development of speech and language is the first six months of life. Delay in diagnosis of hearing impairment accounts for a 0.17-month delay in receptive language development and a 0.30-month delay in expressive language development (21). Newborn screening enables the identification of hearing impairment before 3 months of age, thereby enabling early intervention before 6 months of age (22).

Otoacoustic emission (OAE) is used for newborn screening for congenital hearing impairment. OAE includes a mechanism for sound emission as well as detection of waveforms up to the level of the cochlear. Optimal timing is after the completion of 24 hours of age but can be done as close to 24 hours as possible if indicated. If the baby does not pass the test when done before 24 hours of age, it should be repeated after 24 hours of age to minimize false positives. All newborns should be screened for risk factors for hearing impairment by the medical officer at the same time as the neonatal examination that is conducted within the first 24 hours of life. Risk factors for congenital hearing impairment are given below in table 4.

Table 4: Risk factors for congenital hearing impairment*

1.	Admission to a NICU for more than 7 days
2.	Bilirubin levels at or above exchange levels
3.	Aminoglycoside (gentamicin) administration for more than 5 days (WHO) or loop diuretics (furosemide) or maternal cisplatin therapy
4.	Hypoxic ischemic encephalopathy grade 2 or 3
5.	In utero infections such as cytomegalovirus, herpes, rubella, syphilis and toxoplasmosis, HIV, Hepatitis B, measles, mumps, rubella
6.	Craniofacial malformations (pinna, ear canal, ear tags, ear pits, temporal bone anomalies) or syndromes associated with hearing impairment (Eg. Waardenburg, Alport, Pendred, Jervell and Lange-Nielson, Down, Hunter, Usher, Friedreich ataxia, Charcot-Marie-Tooth, Neurofibromatosis type I and II, Osteopetrosis)
7.	Head trauma / birth trauma (Eg, basal skull (cochlea / middle ear damage) or temporal bone (inner ear) fractures or perforation of the tympanic membrane, bleeding or disruption of the ossicular chain
8.	Meningitis / Culture positive sepsis
9.	Family history of hearing loss in childhood
10.	Prematurity less than 34 weeks gestation

**(adapted with permission from WHO: Universal newborn screening implementation guidance for Southeast Asia and 23.12.2019 Ministry of Health circular on newborn screening for congenital deafness)*

These risk factors increase the risk of the auditory nerve being affected. OAE is only able to detect hearing impairment until the level of the cochlear and does not detect hearing impairment due to the auditory nerve, which is beyond the level of the cochlear. Hence, all newborns with these risk factors mentioned in table 3 are referred to the nearest hospital with audiology and consultant ENT services for hearing screening with automated auditory brainstem response (AABR).

Babies who are born before 34 weeks of gestation are screened once they are medically stable and they reach the corrected gestational age of 34 weeks. This is to minimize false positive results, as the hearing threshold decreases with gestation from 28dB at 28 weeks corrected gestational age to 13dB at 42 weeks corrected gestational age, by about 1 dB/week (5,23,24).

Babies without risk factors are screened using OAE at the bedside, by a nursing officer or an audiology technician, when the baby is quiet, preferably after a feed. Background noise should be minimized by switching off the television / radio in the ward, as well as requesting the staff to be quiet. OAE should be performed away from other electrical devices to minimize electrical interference. Swaddling the baby helps to minimize body movements while reducing the chance of accidental removal of the ear probe. The newborn's ear canal should be visualized for patency. The ear canal should be cleaned with a gauze wick if wax, blood or debris are noted. A different ear probe is used for each baby during each session, where all probes are cleaned at the end of each session. The probe with the best fit for each baby's ear canal is selected and slid all the way through until it can go no further. Once the probe is in position, the clip on the probe cord is fixed on to the newborn's clothes to ensure that the probe remains in place during the test. Holding the probe during the test should be avoided as it blocks the signal. Once the test is started, it will check the probe position and then indicate if the baby passed the test by displaying 'pass' on the screen. Test is repeated in the other ear. Results are recorded separately for each ear.

Screening for hearing impairment is conducted in 3 stages. The first stage and second stage screening are done in the delivery hospital using OAE. First stage screening is done prior to discharge, this can be repeated prior to discharge in those who do not pass the first OAE. Second stage screening is done two weeks after birth, for newborns who do not pass the first stage screening. Second stage screening aims to exclude the transient causes collapse of the ear canal, debris in the ear canal and amniotic fluid or mucus in the middle ear that is more likely to be present soon after birth and are expected to resolve by 2 weeks of age. Neonates who do not pass the second stage screening are referred to the nearest hospital with audiology services and an ENT surgeon, where third stage screening is conducted using automated auditory brainstem response (AABR). Babies who do not pass the third stage screening will receive diagnostic testing with auditory brainstem response (ABR) to confirm the hearing impairment after which the baby will receive speech and language therapy and a trial of hearing aids. Poor response to hearing aids will indicate a cochlear implant.

If the newborn passes the screening test in both ears, at any time point, the newborn is considered to have passed the test. Since OAE screening may not detect a hearing impairment due to an issue with the auditory nerve, parents are advised to continue to monitor the child using the speech and hearing developmental milestones according to the child health development record.

Figure 4: The three-stage newborn screening for congenital hearing impairment *(adapted with permission from the WHO Universal newborn screening implementation guidance for Southeast Asia)*

While the above-mentioned newborn screening programs are in effect at present, screening for eye abnormalities was introduced as part of the pilot project conducted in 2024. This streamlined the examination for eye abnormalities using the naked eye and the ophthalmoscope that is already a part of the neonatal examination. It introduced torch light examination of the eye to detect abnormalities in the eyeball (Anophthalmus), eye lid (infection, hemangioma, coloboma, ptosis),

conjunctiva (infection), cornea (large hazy cornea in glaucoma, megalo and micro cornea), iris (coloboma) and lens (cataract). It also streamlined ophthalmoscopic examination by raising awareness about abnormalities like diminished red reflex and the presence of dark shadows (silhouettes) in the red reflex in addition to detection of the absent red reflex or the presence of the white reflex.

Screening for transcutaneous bilirubin is likely to be the next step in newborn screening in Sri Lanka, as per WHO implementation guidance for Southeast Asia.

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Case Report

A case of neonatal purpura fulminans successfully treated, saving life and limb

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Keywords: Neonatal purpura fulminans; Protein C; Protein S; Thrombosis; Amputation

Introduction

Neonatal purpura fulminans is a rare, life-threatening disease characterised by microvascular thrombosis and disseminated intravascular coagulation (DIC) leading to widespread skin necrosis, circulatory collapse and multiorgan failure[1]. It is caused by inherited deficiencies of proteins C or S and infections. Neonatal purpura fulminans is often fatal, with case fatality as high as 50%[2]. Even among survivors, limb amputation is frequently required. Here, we report a Sri Lankan neonate with severe purpura fulminans successfully treated without limb amputation.

Case presentation

A baby girl was admitted to the neonatal intensive care unit 3 hours after birth due to bluish discolouration of both feet (Figure 1). She was born via elective caesarean section at 38 weeks of gestation with a birth weight of 2.725kg as the second child of previously healthy, non-consanguineous Sri Lankan parents. The antenatal period was uncomplicated, and there were no maternal risk factors for sepsis. There was no family history of thrombophilic or bleeding disorders. The baby had cried at birth, and the Apgar scores were 9, 9, and 10 at 1, 5, and 10 minutes, respectively.

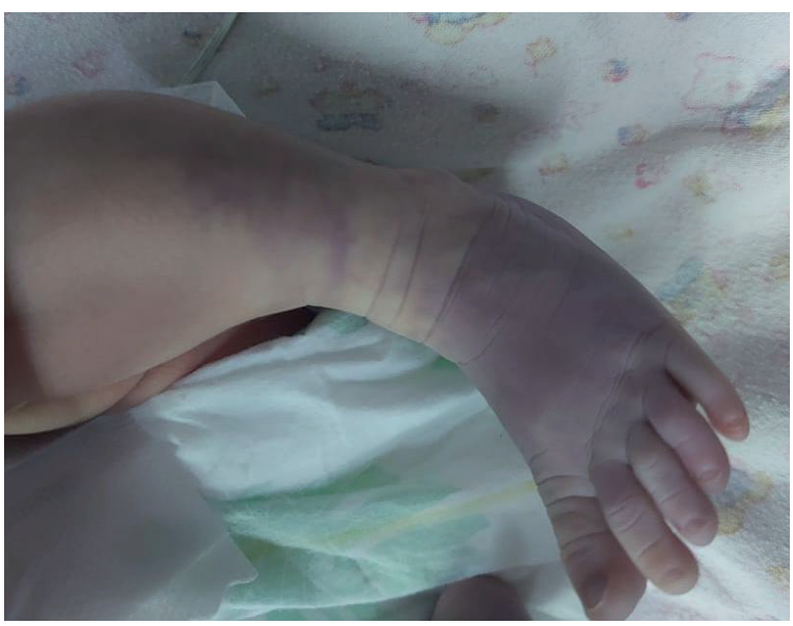

Figure 1- Neonate's foot on admission showing mild bluish discolouration

On examination, the baby was centrally pink and did not have respiratory distress or dysmorphic features. The bluish discolouration was noted in both feet, with evidence of gangrene (Figure 2). Both femoral pulses were palpable, and a grade 2 systolic murmur was heard in the pulmonary area. Her heart rate was 158/min, and the mean arterial pressure was normal (50 mmHg). The oxygen saturation on room air was 88% in both lower limbs and 100% in the right upper limb.

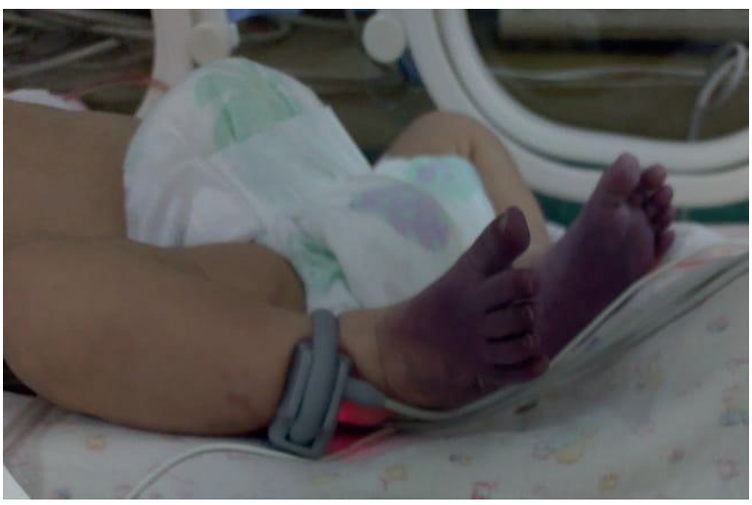

Figure 2- Neonate's feet 3 hours after admission showing extensive discolouration with evidence of gangrene

The initial full blood count showed normal haemoglobin (15.8g/dL), leucocytosis (white blood cell count- 27.7×10^9/L) and thrombocytopenia (platelet count- 77×10^9/L). C-reactive protein was <0.5mg/L. The coagulation profile revealed prothrombin time- 23s (normal 12-16s) and activated partial thromboplastin time- 28s (normal 36-44s). Based on the clinical presentation with lower limb gangrene and thrombocytopenia, a provisional diagnosis of neonatal purpura fulminans was made.

The baby was started on 2L/min nasal prongs oxygen and intravenous penicillin and cefotaxime after obtaining blood for culture. 10 mL/kg platelet-rich plasma, 10 mL/kg fresh frozen plasma (FFP) and 1 unit of cryoprecipitate were transfused. Urgent haematology, vascular surgical, and cardiology referrals were made. Subcutaneous heparin 18 IU/kg was started as the anticoagulant therapy, which was changed to subcutaneous enoxaparin 1 mg/kg twice daily on day two. Intravenous vitamin K 1mg daily was given for five days. FFP transfusions of 10 mL/kg were continued twice daily. Activated protein C concentrates were not available.

The subsequent investigations revealed very high D-dimers (>10,000ng/mL), and the blood picture showed evidence of microangiopathic haemolytic anaemia. Blood cultures were sterile. Doppler studies of both lower limbs, echocardiography and abdominal ultrasonography showed normal results. Protein C and S levels were normal; however, they were done after FFP transfusions.

On day five, the baby developed circulatory failure, fever and tonic-clonic seizures. She was intubated and ventilated and commenced on three inotropes (dopamine, dobutamine and noradrenaline), and the antibiotics regimen was upgraded to intravenous meropenem and vancomycin. Seizures were treated with intravenous phenobarbitone and levetiracetam. Her blood glucose, electrolytes, calcium,

magnesium, electroencephalogram and cranial ultrasonography were normal.

By day ten of life, the baby's condition improved, and the necrotic areas were limited to the superficial aspects of the affected toes (Figure 3). Twice daily FFP administration was continued until day 24, which was thereafter given daily until day 31, every other day for the next four weeks, and weekly until three months. Subcutaneous enoxaparin was tailed off gradually. Surgical amputation was not required, and all affected toes were salvaged except for the distal phalanx of the right third toe, which underwent auto amputation (Figure 4).

At the 3-month review, the baby had normal weight for her age, was developmentally age-appropriate and had well-healed lower limbs. She is awaiting repeat testing for protein C and S levels at six months.

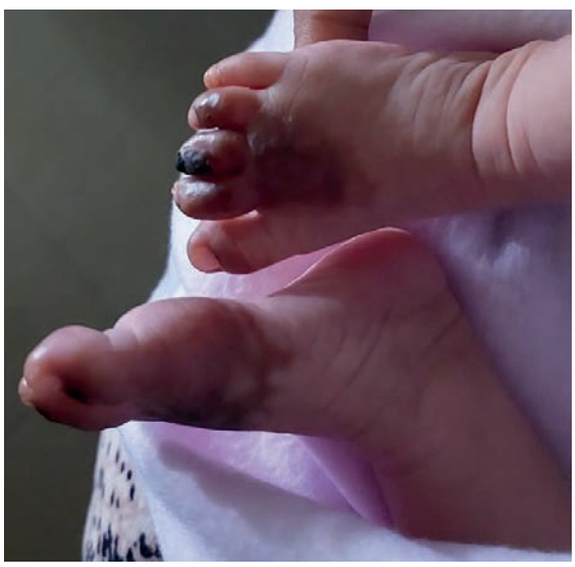

Figure 3 - Baby's feet at day 10 showing necrotic areas limited to the superficial aspects of the right third toe

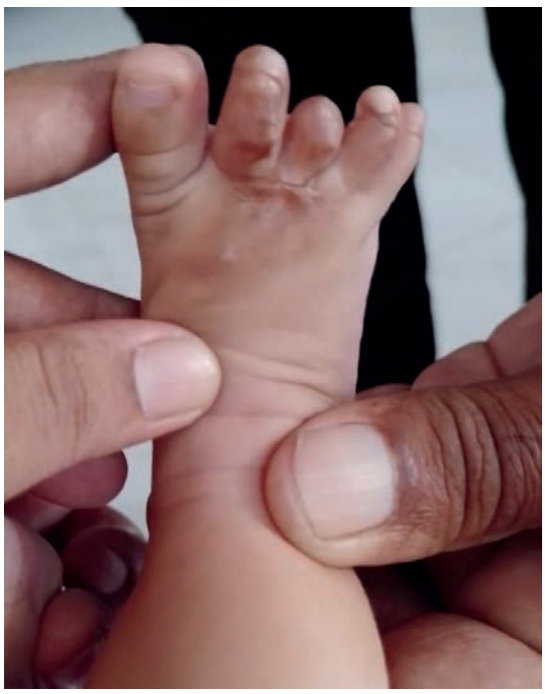

Figure 4 - Baby's feet at three months showing complete healing with auto amputation of the distal phalanx of the right third toe

Discussion

Neonatal purpura fulminans is a severe and fatal condition caused by occlusion of small and medium-sized blood vessels due to microvascular thrombi. It has both inherited and acquired forms. Inherited neonatal purpura fulminans is caused by protein C or protein S deficiency. In contrast, infections leading to consumptive coagulopathy due to DIC coupled with a relative deficiency of protein C is the cause of the acquired form of purpura fulminans[1]. Proteins C and S are synthesised in the liver, and their deficiencies lower the ability to decrease thrombin production, leading to a hypercoagulable state[3].

Inherited neonatal purpura fulminans are due to genetically inherited homozygous or compound heterozygous mutations in the genes encoding protein C or protein S[4]. If a

family history is positive for protein C or S deficiency or there is a history of thrombosis in the family, prenatal diagnosis by chorionic villous sampling is recommended to detect genetic mutations of protein C or protein S. Antenatal diagnosis of protein C deficiency offers significant benefits by providing an opportunity to prevent complications of the disease. The clinician can deliver the neonate early and commence replacement therapy as early as possible[4].

The most identified pathogen causing acquired neonatal purpura fulminans is Group B Streptococcus (GBS)[1]. In addition, it is caused by *Escherichia coli*, varicella, meningococcus, pneumococcus, *Klebsiella oxytoca*, *Neisseria meningitidis* and *Citrobacter*. There was also evidence that *Acinetobacter baumannii* is another potential causative agent[5].

Neonates with purpura fulminans should receive treatment with anticoagulant medications promptly[6]. The mainstay of therapy includes FFP transfusion, anticoagulation and protein C replacement. Even with the optimal management of protein C concentrates, the mortality is high, and a significant proportion of survivors have permanent disability due to limb amputation[7]. A case series of 16 children reported that 69% of survivors required amputation[8].

The most important feature of this case report is that the neonate survived without limb amputation.

She was diagnosed within the first 24 hours of life and was successfully treated, saving life and limb despite being in a resource-limited setting without protein C concentrates. Due to the unavailability of protein C concentrates, the patient was managed only with FFP transfusion, anticoagulation and antibiotics. We believe that the prompt diagnosis and timely treatment with available therapeutic modalities resulted in a near-complete recovery of the baby.

Screening for parents' protein C and S levels did not support a diagnosis of inherited protein C deficiency in our patient. All cultures were sterile; however, c-reactive protein was high during the first week, and the clinical picture suggested a severe infection. Rapid response to upgraded antibiotics supported the presumptive diagnosis of acquired purpura fulminans secondary to severe infection in the neonate.

In conclusion, our case report highlights the importance of the prompt diagnosis of neonatal purpura fulminans to commence early and appropriate treatment. It also shows that neonatal purpura fulminans can be effectively treated with FFP as a replacement for protein C, alongside anticoagulation and an appropriate combination of antibiotics for an extended period. This evidence-based management approach can succeed in low-resource settings, saving lives and not necessitating amputation.

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Picture Story

An unusual presentation of congenital haemangioma as hypertrophy of the hand at birth

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Keywords: Congenital haemangioma; Vascular tumours; Hypertrophy of hand; Haemangioma.

Background

Congenital haemangiomas are rare, benign vascular tumours that are present in their maximum sizes at birth [1]. In contrast to the common form of infantile haemangioma, they do not grow postnatally and a majority regress over the first few months [2]. Congenital haemangiomas usually appear as well-demarcated red-purple plaques or exophytic masses that grow outward in the skin [3]. Involvement of the deep subcutaneous tissue or muscles is rare. Here, we report a newborn with a congenital haemangioma presenting as a large soft tissue mass causing hypertrophy of the right hand.

Case presentation

A baby girl was born at term via emergency caesarean section due to meconium-stained liquor. She was the third child of healthy, non-consanguineous, Sri Lankan parents. **Antenatal ultrasound scans did not reveal any abnormality.** The baby weighed 2.4kg at birth and had Apgar of 9, 10 and 10 at 1, 5 and 10 minutes, respectively. Gross enlargement and bluish discolouration of the right hand were noted at birth.

On examination, the baby was active, alert, pink and did not have respiratory distress. Marked hypertrophy of the right upper limb involving the lower forearm, wrist, hand and all fingers was noted (Figure 1). The length and width of the right hand were 10.8cm and 7.6cm, respectively, while they were 6.5cm and 3.2cm in the left hand. All peripheral pulses, including the right radial pulse, were palpable, and the heart rate was 120/minute. There were no haemangiomas or vascular malformations in other body parts, and the eye examination was normal. Oxygen saturation of both upper limbs on air was 100%. A clinical diagnosis of congenital haemangioma involving the right upper limb was made.

Figure 1 – Marked hypertrophy of the right hand and fingers

Her full blood count, c-reactive protein, prothrombin time, and activated partial thromboplastin time were normal. The x-ray showed soft tissue swelling in the right hand with normal bone anatomy; there was no evidence of bone hypertrophy, erosions or focal abnormalities (Figure 2). The ultrasound of the right hand showed a soft tissue thickening with abnormal blood vessels in the palmer and dorsal aspects of the right hand, confirming the diagnosis of congenital haemangioma. Ultrasound scans of the brain, abdomen and kidneys were normal.

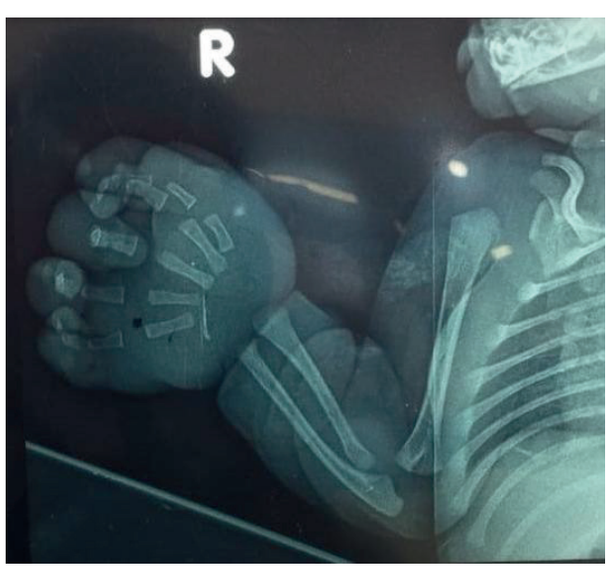

Figure 2 – X-ray of the right hand showing soft tissue swelling with normal bony anatomy

The baby was referred to plastic and vascular surgical teams, and they agreed on a decision to manage conservatively, expecting a spontaneous resolution. At the 2-month review, the right-hand hypertrophy showed nearly 50% spontaneous regression (Figure 3). All movements of the small joints of the right hand were normal.

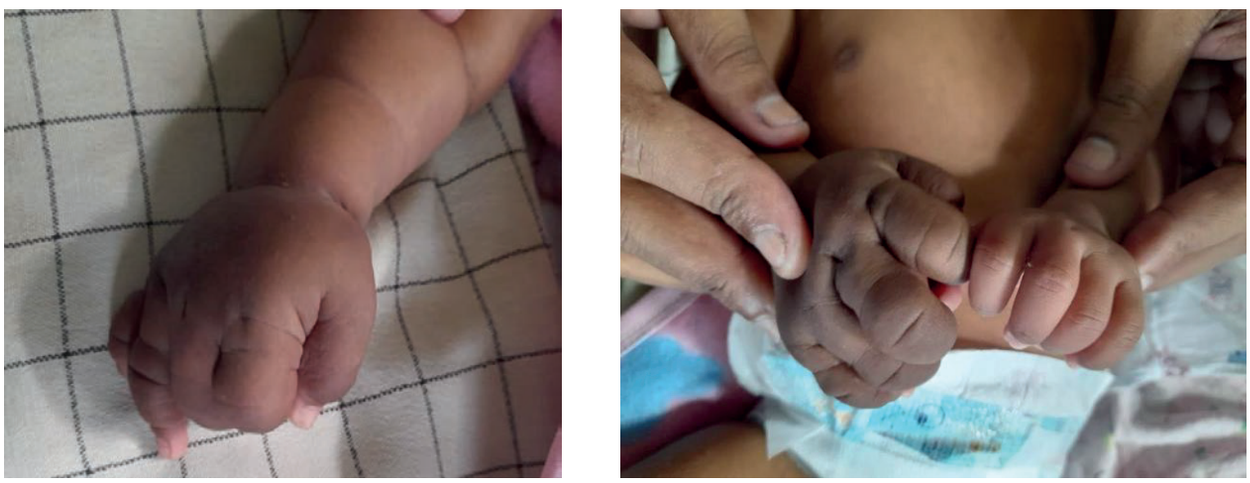

Figure 3 – Right hand at two months showing spontaneous resolution

Discussion

Vascular lesions in newborns are broadly divided into vascular tumours and vascular malformations [4]. Vascular tumours can be benign or malignant, whereas vascular malformations are classified based on their tissue origin. Vascular malformations are present at birth, become more apparent over time, and expand in proportion to the infant's overall growth but do not grow out of proportion to body growth. In contrast, benign vascular tumours classically show growth out of proportion to body growth and involute with time. The most common form of benign vascular tumour is infantile haemangiomas, which are not present or are slightly evident at birth and rapidly grow during the first months of life, followed by a gradual regression during the pre-school years. Congenital haemangiomas are a rare subtype that present at their full sizes at birth and involute over the first few months of life [1]. The vascular lesion in our baby was present at birth at its full size and showed involution over time, confirming the diagnosis of congenital haemangiomas.

The typical clinical presentation of congenital haemangioma is a plaque-like or exophytic lesion of a few centimetres appearing on the surface of the skin [3]. The neonate in our case report had vascular lesions involving the soft tissue of the palmer and dorsal aspects, resembling massive hypertrophy of the right hand at birth. Therefore, our case report describes a very rare presentation of congenital haemangioma.

The management of congenital haemangioma depends on the size and location of the lesion. Conservative management with careful observation is the preferred mode of treatment in most congenital haemangiomas that are noncomplicated and do not impair function [5]. Oral propranolol, a commonly used medical treatment for infantile haemangioma, is not recommended for congenital haemangioma [6]. Due to the rapid involution and involvement of deeper structures, our patient was managed conservatively with observation. At the two-

month review, the child showed remarkable improvement, with regain of functionality of the right hand.

In conclusion, we described a rare congenital haemangioma presenting as a localised hypertrophy of the right hand in a newborn. The case report highlights the value of making the accurate clinical diagnosis of congenital haemangioma instead of common infantile haemangioma to decide on the appropriate management for a better overall outcome for affected neonates.

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