NATIONAL PROTOCOL FOR SCREENING OF CRITICAL CONGENITAL HEART DISEASES OF NEWBORNS BY PULSE OXIMETRY

Ministry of Health
Republic of Maldives

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FOREWORD

It gives me real pleasure to present the National Protocol for screening of Critical Congenital Heart Disease in Newborns. The development of this National Protocol is a major step in the efforts to end preventable newborn deaths and to guide the efforts for accelerating work towards the long-term goal of ending preventable deaths among children.

There have been significant progress made in recent decades in reducing the number of child deaths, but neonatal mortality rate has declined at a slower pace, birth defects causing significant morbidity and mortality. Further, Congenital heart disease in turns leads to a huge medical burden, economic burden and mental burden for the family and for society. Therefore, in order to further reduce child mortality and end preventable deaths, intensified action and guidance are needed to ensure newborn survival. Also early detection of CHD would improve the outcome through timely appropriate treatment

This Protocol provides a national guideline on implementing pulse oximetry screening for CCHD. National Protocol for screening of Critical Congenital Heart Disease in Newborns is based on a review of latest literature as well as the current best-practice on the subject.

Pulse oximetry being a safe, noninvasive, inexpensive, and reasonably sensitive test is proven to have detected many cases of Critical Congenital

heart disease (CCHD). Congenital Heart Disease which has been considered as the most common cause of infant death worldwide and is true for Maldives.

I wish to thank the many stakeholders, professionals, experts and organizations who have provided input into the development of this protocol.

Abdulla Nazim Ibrahim
Minister of Health

PREFACE

Congenital Heart Disease (CHD) is a medical condition that can afflict any child, cutting across racial and socioeconomic differences in the community. CHDs are the most common types of birth defects, worldwide, including the Maldives. CHD accounts for 30% of deaths in children with birth defects. CHD impacts the wellbeing of the child as well as the entire family. Therefore, early detection of the condition is vital in the provision of appropriate care for the afflicted child.

Universal screening of babies after 24 hours of birth using pulse oximetry is a proven method for early detection of congenital heart disease. Pulse oximetry is a tool that measures oxygen saturation and based on the presence of hypoxemia, it can assist identifying many cardiac lesions. It is an effective tool for medical professionals in the early diagnosis and treatment of CHD, having the potential to save lives.

We are excited to implement a CHD screening programme nationwide in the Maldives, involving this Protocol for using pulse oximetry screening of newborns. It is based on a review of latest literature on pulse oximetry screening for Critical Congenital Heart Diseases (CCHD) as well as the current best-practice on the subject. This Protocol provides a national guideline on implementing pulse oximetry screening for CCHD.

The existing approach to the subject is based on prenatal testing using ultrasound technology and postnatally, through physical examination or by presentation of symptoms during the first 24 hours of life. However, it has been shown that prenatal testing provides a diagnosis only in 23% of cases of pregnancies while postnatal care, in the absence of appropriate screening, often presents more complications. Pulse oximetry screening targets specific anomalies classified as CCHD. Failure to detect such heart defects within the

first few days or weeks of life may lead to critical events such as cardiogenic shock or even death. Survivors who present late are at greater risk of neurologic injury and subsequent development delay. Many of these conditions often require emergency care involving at times the transfer of very critical newborn to tertiary care centers from the Peripheral Hospitals. The complex challenges presented by our dispersed island-geography add to the complications. Early detection of CCHD can potentially improve prognosis and diminish mortality and morbidity rate of affected infants.

Hence, the national screening of all newborns is a crucial tool for medical practitioners in this country. A normal newborn should have an oxygen saturation of 95% to 97% after 24 hours of life. We have developed an algorithm for CCHD Screening that will be followed through as part of the implementation of this Protocol. If the infant should fail their CCHD screening by pulse oximeter, our policy will be to notify a pediatrician and all future decisions relating to care of newborns with lower than expected oxygen saturations, will be made at the discretion of the pediatrician or pediatric cardiologist caring for the infant. It is recommended that a physical exam be done along with obtaining an echocardiogram to rule out structural abnormalities for newborns with abnormal pulse oximetry readings.

This Protocol is the culmination of work by many stakeholders, professionals, experts and organizations. We would like to express our gratitude to the Ministry of Health for their support in launching this initiative. It is made possible by the help of the Head of Department of Child Health, Dr. Niyasha Ibrahim. We would also like to thank WHO and UNICEF for their assistance and Department of Obstetrics and Gynecology for their back up in its implementation. We extend our appreciation to the healthcare providers and the NGOs for their arduous efforts in working with us in this field.

We request that you continue with your good work by helping us in the national implementation of this CCHD Screening using pulse oximetry. It is an historic opportunity to step in and potentially save the lives of precious children, and improve their quality of life and lifespan. Let us detect CCHD early. Let us work together in giving these dear and loved hearts a chance to live a better life before they leave hospital.

May Allah Bless all the good work you are doing, every day – Amen.

Sincerely,

Aishath Eleena *MD (pediatrics),*

Fellowship in Pediatric Cardiology (Thailand), Consultant Pediatric Cardiologist

Indira Gandhi Memorial Hospital (IGMH)

IS PULSE OXIMETRY SCREENING RECOMMENDED?

Universal screening of babies after 24 hours of birth using pulse oximetry is a proven method for early detection of congenital heart disease. Pulse oximetry screening for CCHD and is endorsed by many eminent advocates such as:

- American Heart Association
- American Academy of Pediatrics
- American College of Cardiology
- American College of medical genetics

Pulse oximetry screening of all newborns is also being used by an increasing number of hospitals in many countries all over the world, many having issued national guidelines recommending universal screening.

WHY IS THIS SCREENING IMPORTANT?

Studies from the United States and other developed countries have shown that as many as 30 to 50% of infants with CCHD are discharged after birth without being identified**.** This diagnostic gap is likely to be even higher in low resource countries including Maldives. CHD accounts for 30% of deaths

in children with birth defects. Surgical and catheter interventions now lead to excellent outcomes for most cases of CCHD. Therefore, early and timely detection of these conditions are vital in the provision of appropriate care for the afflicted child.

SOME FACTS ABOUT CONGENITAL HEART DISEASE (CHD)

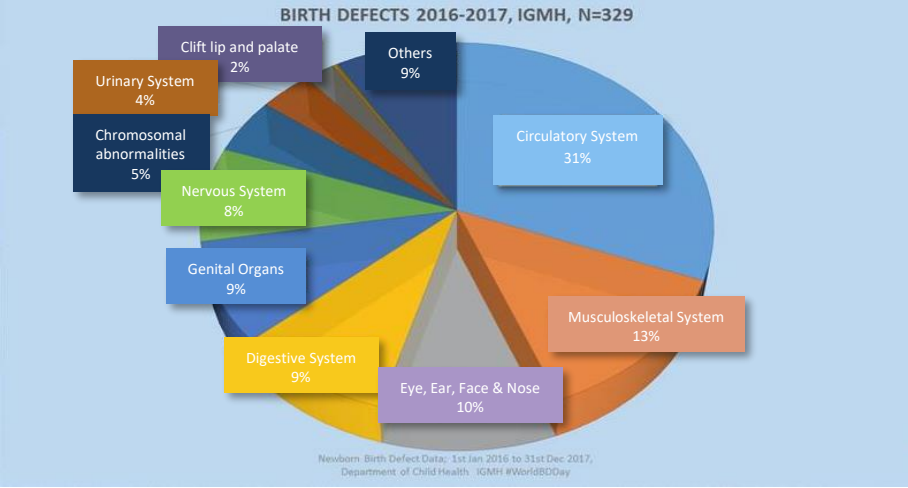
- The most common of all birth defects , worldwide, including the Maldives
- Affects 1 in 100 babies
- Accounts for 30% of all infant deaths due to birth defects

SOME FACTS ABOUT CRITICAL CONGENITAL HEART DISEASE (CCHD)

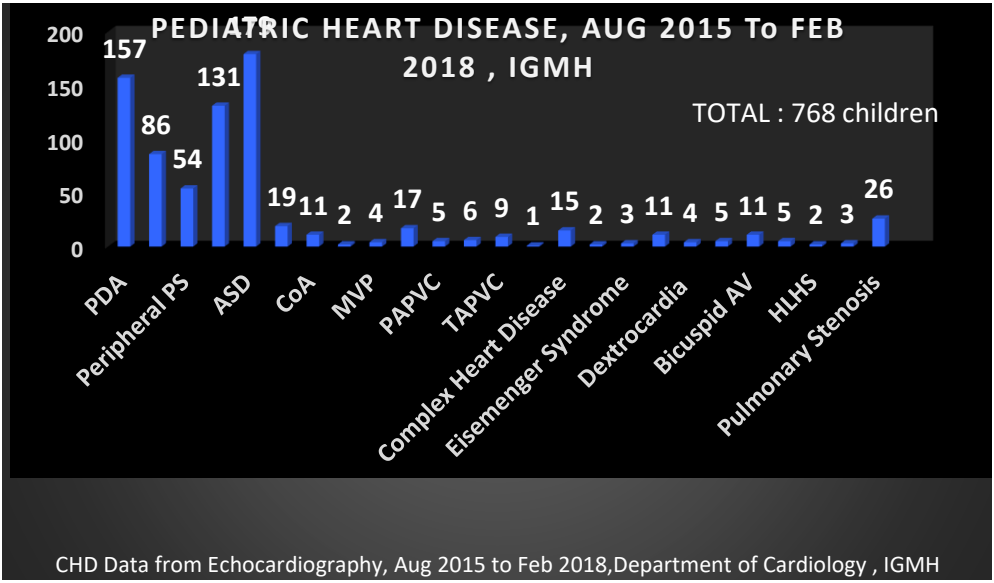
- CCHD is a heart lesion for which neonates require surgery or catheter-based intervention, soon after birth to survive
- 17- 33% of CHD is actually CCHD
- CCHD can often be identified by prenatal USG : HOWEVER
- 50% of all infants with CCHD are discovered after the infant is discharged from the hospital

INCIDENCE OF CONGENITAL HEART DISEASE IN MALDIVES

In the Birth Defect Registry (WHO – SEARO newborn birth defect Database) congenital heart disease is registered under the category congenital malformations of the circulatory system which is from Q20 to Q28. In the year 2016 and 2017 there were 329 children with birth defects in IGMH, out of which the major portion, 31%, comprised of children with congenital heart diseases as shown in the pie chart below.



Looking at the data from IGMH, Echocardiography lab (shown below in a bar graph) we also know that there are a significant number of children with heart diseases in the Maldives.



LET US PREVENT THIS

- Unfortunately, a seemingly normal, healthy infant can suddenly experience serious or life-threatening complications within the first few days or weeks of life that requires emergency care.
- According to a study done in California, the median age of death due to undiagnosed CCHD is <2weeks of age.

HOW CAN PULSE OXIMETRY TESTING HELP

- Detection of CCHD increases to about 85% before hospital discharge with the use of Pulse Oximetry as part of an infant's routine newborn screen.
- With the use of Pulse Oximetry, CCHD benefits from early detection and interventions such as
    - ✓ Surgery
    - ✓ Heart catheterization
    - ✓ Medications

Congenital heart disease lesions divided by likelihood of being detected by pulse oximetry

Primary Targets	Secondary Targets	Possibly Screenable	Not Screenable
LHS	Interrupted aortic arch/aortic atresia	Aortic stenosis with PDA	Coarctation of the aorta without a PDA
TOF	Coarctation of the aorta with PDA	Pulmonary stenosis	Ebstein's anomaly without right-to-left shunt
Pulmonary Atresia	Ebstein's anomaly	Complete atrioventricular canal	Aortic stenosis without PDA
Tricuspid Atresia	Double-outlet right ventricle		Other left-to-right shunting lesions
TGA	Single ventricle physiology		
TAPVC			
Truncus Arteriosus			

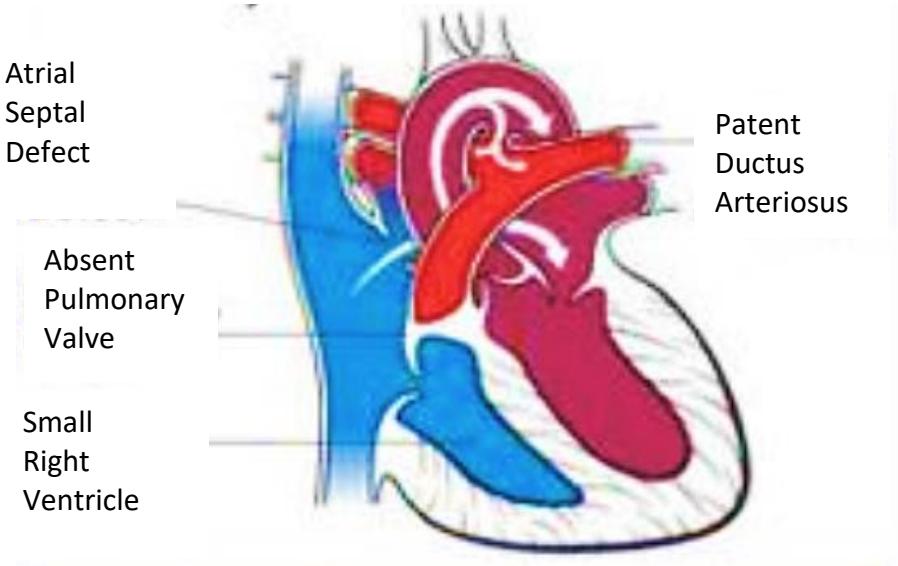
THE SEVEN TARGET CCHD LESIONS

Pulse oximetry screening targets specific anomalies classified as CCHD. The main seven targeted lesions which can be easily detected by pulse oximetry screening are listed below and elaborated.

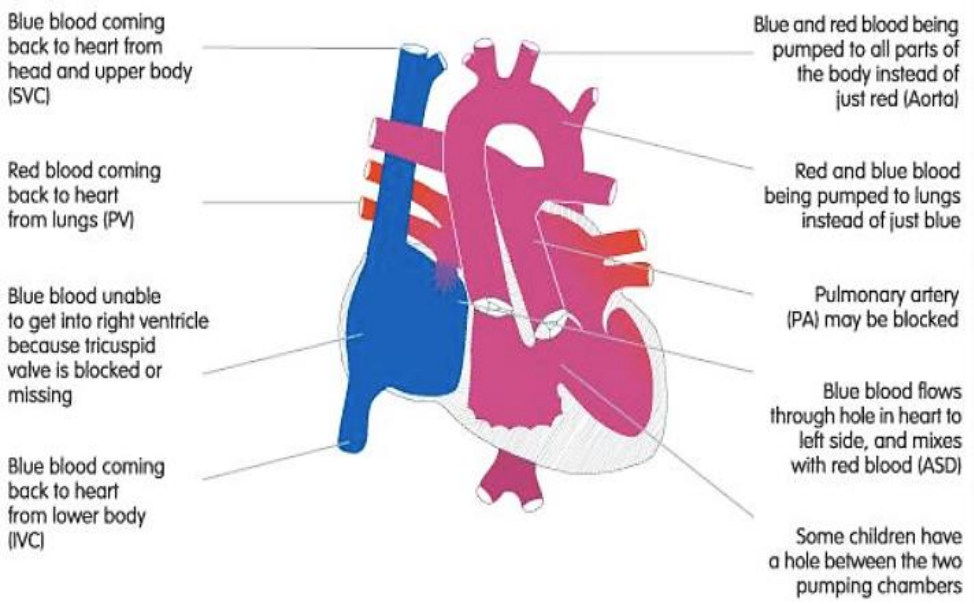
- Pulmonary atresia
- Tricuspid Atresia

- Tetrology of Fallot (TOF)
- Total Anomalous Pulmonary Venous Return (TAPVC)
- Transposition of Great Arteries (TGA)
- Truncus Arteriosus
- Hypoplastic Left Heart Syndrome (HLHS)

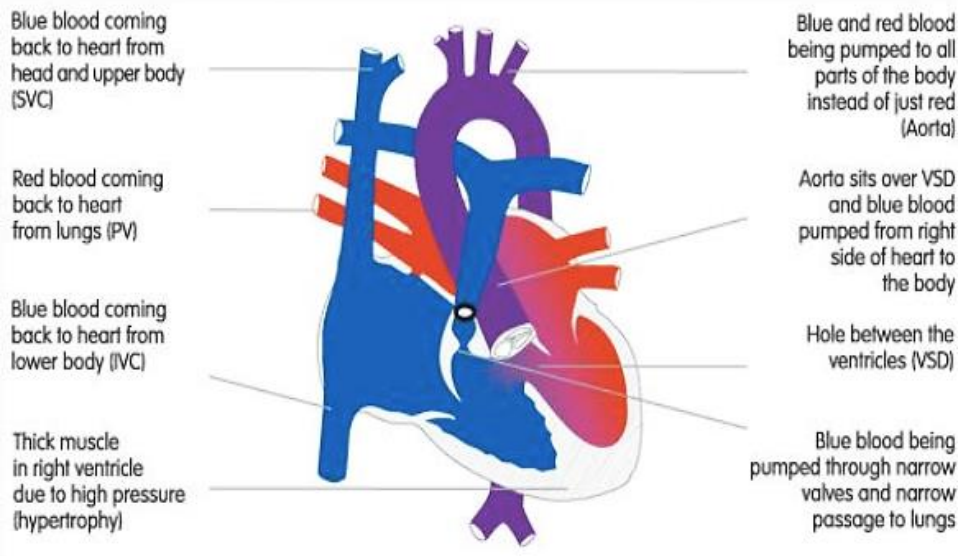
PULMONARY ATRESIA



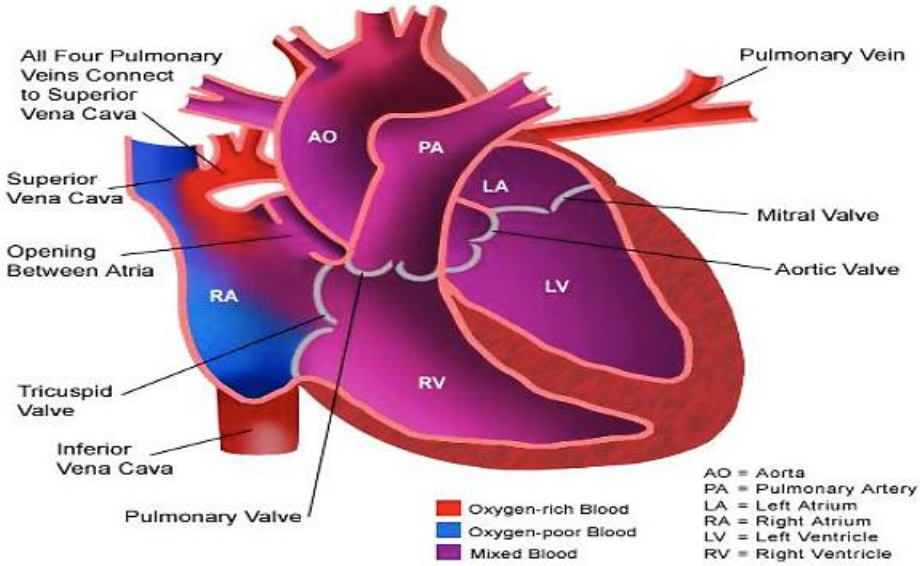
TRICUSPID ATRESIA



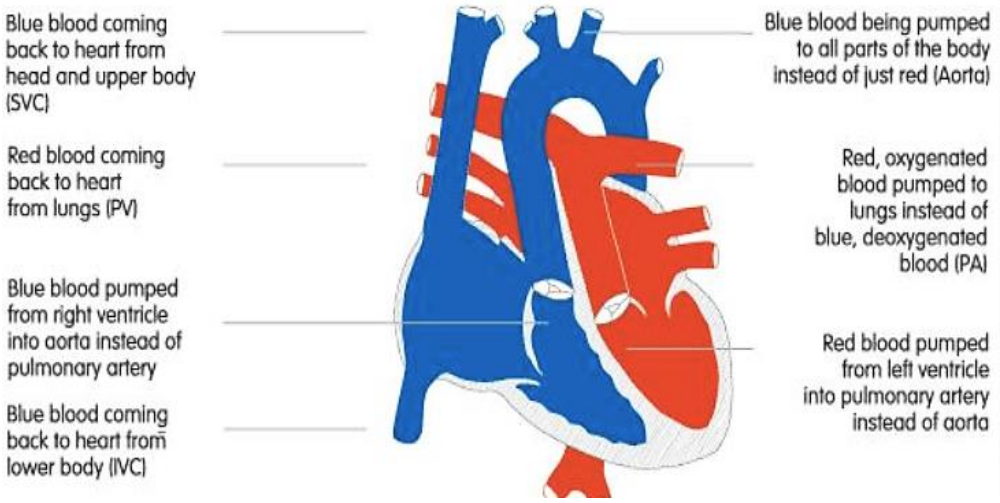
TETRALOGY OF FALLOT (TOF)



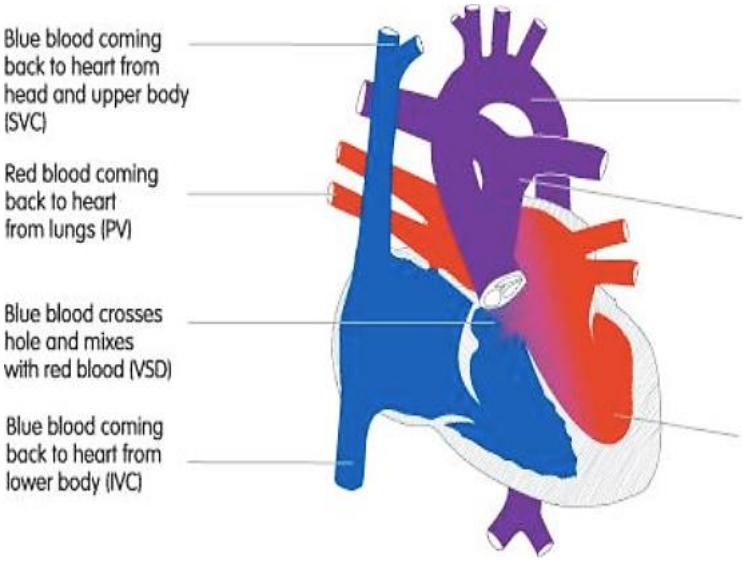
TOTAL ANOMALOUS PULMONARY VENOUS RETURN (TAPVR)



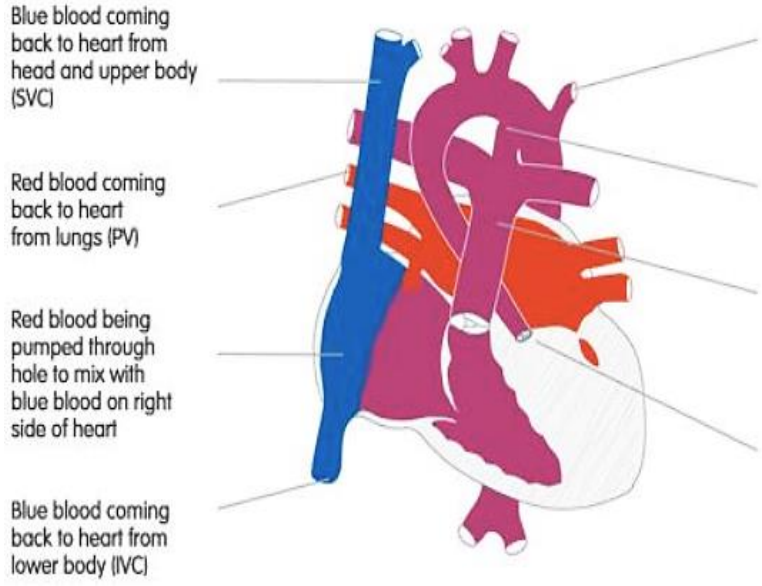
TRANSPOSITON OF GREAT ARTERIES (TGA)



TRUNCUS ARTERIOSUS



HYPOPLASTIC LEFT HEART SYNDROME (HLHS)



WHO SHOULD BE SCREENED?

- Newborns who are 24-48 hours of age. The closer to 48 hours, the better.
- If an early discharge is planned, Pulse Oximetry screening should be performed as late as possible before hospital discharge.
- Infants in neonatal intensive care units should be screened at 24-48 hours of age or when medically appropriate after 24 hours of age.

OVERVIEW OF PULSE OXIMETRY SCREENING

- Pulse Oximetry:
  - Measures the oxygen saturation of the blood
  - Is non-invasive and painless
  - Is accurate and reliable
  - Is fast and easy to perform
  - Is inexpensive
- Pulse Oximetry testing should always be done in room air and should be used as a compliment to the physical examination and should NEVER be used as a replacement
- Contrary to the rest of the newborn screening panel, interpretation of Pulse Oximetry test results and decisions regarding intervention will take place at the hospital and birthing facilities at the actual time the test is performed.

EQUIPMENT USED FOR SCREENING

- Each birthing facility will be responsible for selecting and securing Pulse Oximetry equipment for screening newborn for CCHD, if appropriate equipment is not already available.
- Standard of the Pulse Oximetry equipment must be :
  - Cleared by the FDA for use in newborns
  - Calibrated regularly based on manufacturer guidelines

TIPS ON PROPER SCREENING

- Conduct the screening in a quiet area
- Do not attempt to perform Pulse Oximetry on an infant while the baby is crying or if the baby is cold
- If possible, conduct screening while the infant is awake and calm
- Make sure the infants right hand and both feet are clean and dry Substances , such as dried blood, can affect the pulse ox reading

PULSE OXIMETRY PROBE PLACEMENT

- Choose the proper application site on the outside, fleshy part of the infants hand or foot.
- Place the light emitter portion of the probe on the top of the hand or foot.
- Place the photodetector directly opposite of the light emitter, on the bottom of the hand or foot.

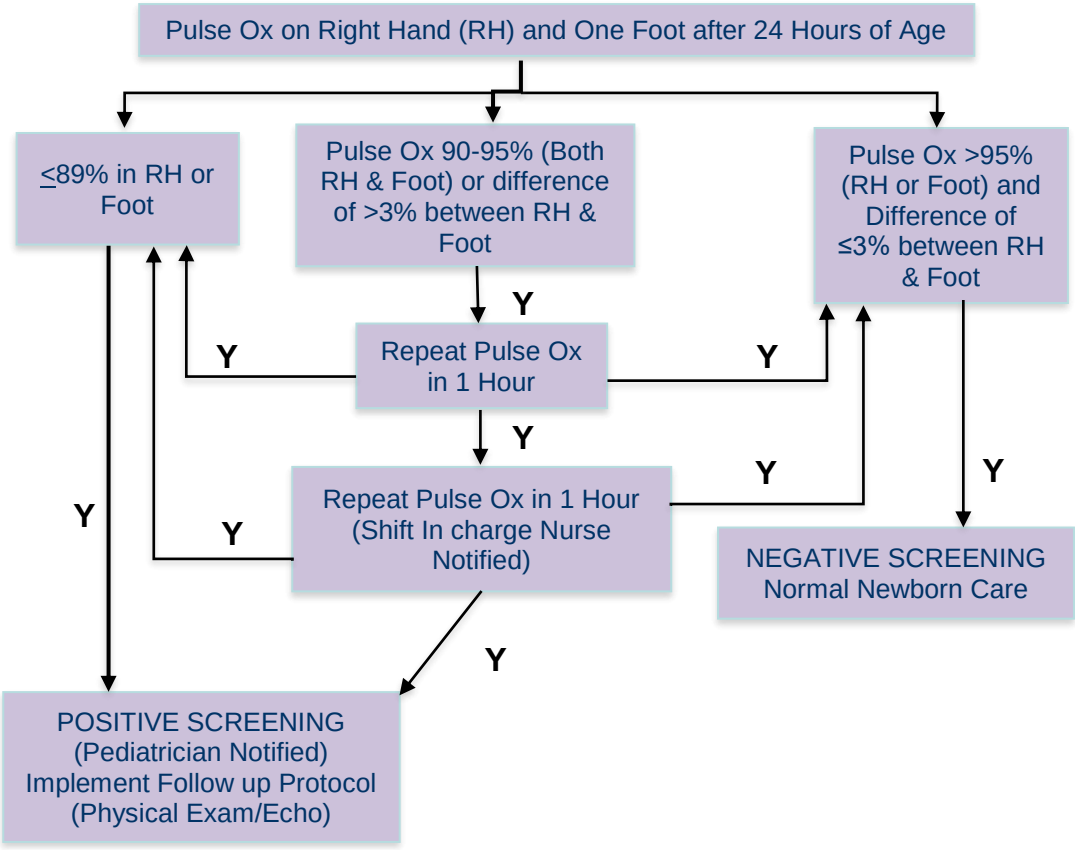
- REMEMBER : The photodetector and the light emitter must be directly opposite each other in order to obtain an accurate reading
- Secure the probe to the infant's hand or foot using the adhesive or form tape recommended by the supplier.

PROPER ALIGNMENT

Application with Disposable

Application with Reusable

CCHD SCREENING PROTOCOL

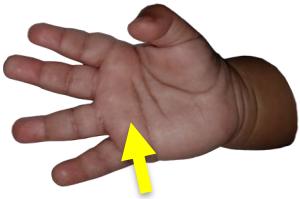


Y= if yes

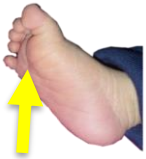
Pulse Ox= Pulse Oximetry

PULSE OXIMETRY SCREENING PROTOCOL FOR ASYMTOMATIC NEWBORNS FOR CRITICAL CONGENITAL HEART DISEASE (ALWAYS DO IN ROOM AIR)

- Check Pulse Oximetry at 24-48 hours of life OR
- Shortly before discharge if <24hours of age OR
- When medically appropriate before discharge if in NICU

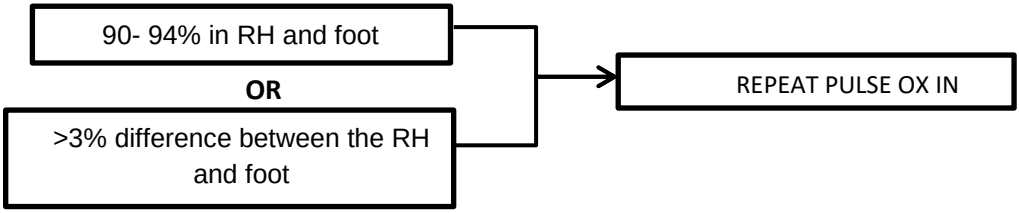


RH Application Site

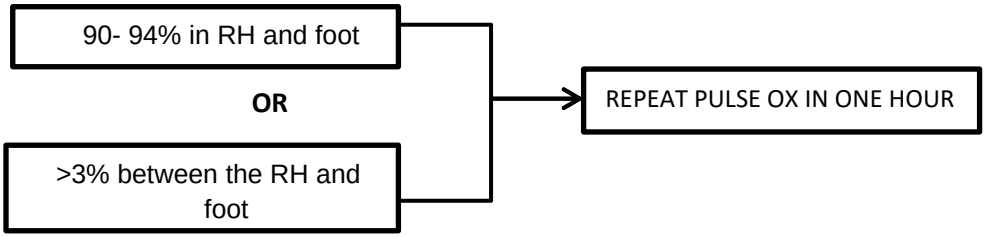


Foot Application Site

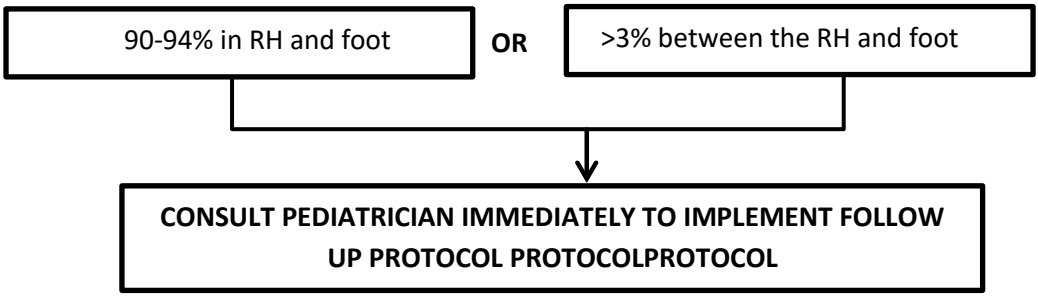
IF THE INITIAL PULSE OX READING IS ABNORMAL



IF THE SECOND PULSE OX READING IS



IF THE THIRD PULSE OX READING IS



FAILED SCREEN AND FOLLOW UP PROTOCOL

- On call Pediatrician Notified
- Pediatrician orders Pediatric Echocardiogram
- Pediatric Echocardiogram must be performed by a trained pediatric sonographer
- Pediatrician consults Pediatric Cardiologist
- Pediatric Cardiologist will determine further diagnostic follow up
- Hospital staff confirms that CCHD Data collection Form (Birth Defect Form) has been filled and sent

DATA ENTRY – CCHD RESULTS

- Same system as data entry for the Birth Defect Registry (WHO – SEARO newborn birth defect Database)
- CCHD tab (Birth Defect category Q20 to Q28 ; Congenital malformations of the circulatory system)
- Should be completed by staff that performs other Birth Defect data entry

CCHD SCREENING INFORMATION

INITIAL SCREEN

- Hospital Number :
- Patient Name :
- Birthing facility : (Hospital Name)
- Date of Birth :
- Time of Birth :
- Gestational age at birth :
- Date of Screening (mm/dd/yyyy)
- Time of Screening (24 hours)
- Age at Screening : (Hours of life)
- SPO2 of Right Hand (RH) : %
- SPO2 of Foot : %
- Difference in oxygen saturation (RH and Foot) : %

- Performed by : Name of Staff
- **PASS FAIL N/A**
- IF FIRST SCREEN IS **"PASS"** SECOND SCREENING IS NOT REQUIRED AND BABY CAN BE DISCHARDED

IF FIRST SCREEN IS **"FAIL" NEED TO DO SECOND SCREEN IN 1 HOUR OR IF THE SPO_2 is <90% in RH OR FOOT NEED TO REFER IMMEDIATELY**

SECOND SCREEN

- Information from previous screening is re-populated
- If second screen is **"PASS"**
  - Third screen is not required
  - No further action is necessary
  - If second screen is a **"FAIL"**
  - Perform third screen in 1 hour

THIRD SCREEN

- Information from previous screening is re-populated
- Data fields are same as initial screen
- If third screen is a **"PASS"**
  - No further action is necessary
- If Third screen is a **"FAIL"**

IMPLEMENT THE FAILED SCREEN FOLLOW UP PROTOCOL (NOTIFY PEDIATRICIAN)

ANOTHER REVIEW OF FAILED SCREEN FOLLOW-UP PROTOCOL

- On call Pediatrician Notified
- Pediatrician orders Pediatric Echocardiogram
- Pediatric Echocardiogram must be performed by a trained pediatric sonographer
- Pediatrician consults Pediatric Cardiologist
- Pediatric Cardiologist will determine further diagnostic follow up
- If the pediatric echocardiogram and clinical assessment indicates other conditions that may affect pulse oximetry (ie : Pulmonary disorders), Cardiologist informs to Pediatrician
- Hospital staff reconfirms that CCHD Data collection Form (Birth Defect Form) has been filled and sent

SCREEN N/A

- When a Pulse Oximetry screen is not performed due to:
  - Prenatal Diagnosis of CHD
  - Transferred to another facility before 24hours
  - Or if another reason need to specify the circumstances

IMPLEMENT THE PULE OXIMETRY SCREENING AND SAVE A LIFE BY EARLY DETECTION OF A BABY WITH CCHD

ANNEX

PLACE LABEL OR WRITE-IN INFORMATION

Patient Name ……..... Hospital Number ……..... Time of birth …… Date of birth …../…./…… Gestational age at birth …….. Birthing Facility Name ……..……

Congenital Heart Disease Screening Program:
Screening Form

Initial Screening: (circle Pass/ Fail or N/A where applicable)

Date of screening (mm/dd/yyyy) …../…../…….
Time of Initial Screening (24 hours) ……..
Age at Initial Screening: (Hours of life) ……..
SPO2 of Right Hand: …….. %
SPO2 of Foot: ……%
Difference in Oxygen saturation (RH and Foot) …… % Pass Fail N/A

Second Screening (1 hour following initial screen if fail initial screen)

Date of Screening (mm/dd/yyyy) …../…../…….
Time of Initial Screening (24 hours) ……..
Age at Initial Screening: (Hours of life) ……..
SPO2 of Right Hand: …….. %
SPO2 of Foot: ……%
Difference in Oxygen saturation (RH and Foot) …… % Pass Fail N/A

Third Screening (1 hour following second screen if fail second screen)

Date of Screening (mm/dd/yyyy) …../…../…….
Time of Initial Screening (24 hours) ……..
Age at Initial Screening: (Hours of life) ……..
SPO2 of Right Hand: …….. %
SPO2 of Foot: ……%
Difference in Oxygen saturation (RH and Foot) …… % Pass Fail N/A

*** If pulse ox saturation is <90% in either the hand or foot the Pediatrician must be notified immediately. "Fail must be checked".**

*** If pulse ox saturations are <95% in both the hand and foot or there is a >3% difference between the two on three measures each separated by one hour the Pediatrician must be notified. "Fail must be checked.**

*** If pulse ox saturations are >95% in either extremity, with a <3% difference between the two readings infant is expected to be normal. "Pass" should be checked".**

Screener's Name ……..

Screener's Signature ……..... Date ……/……. /…….

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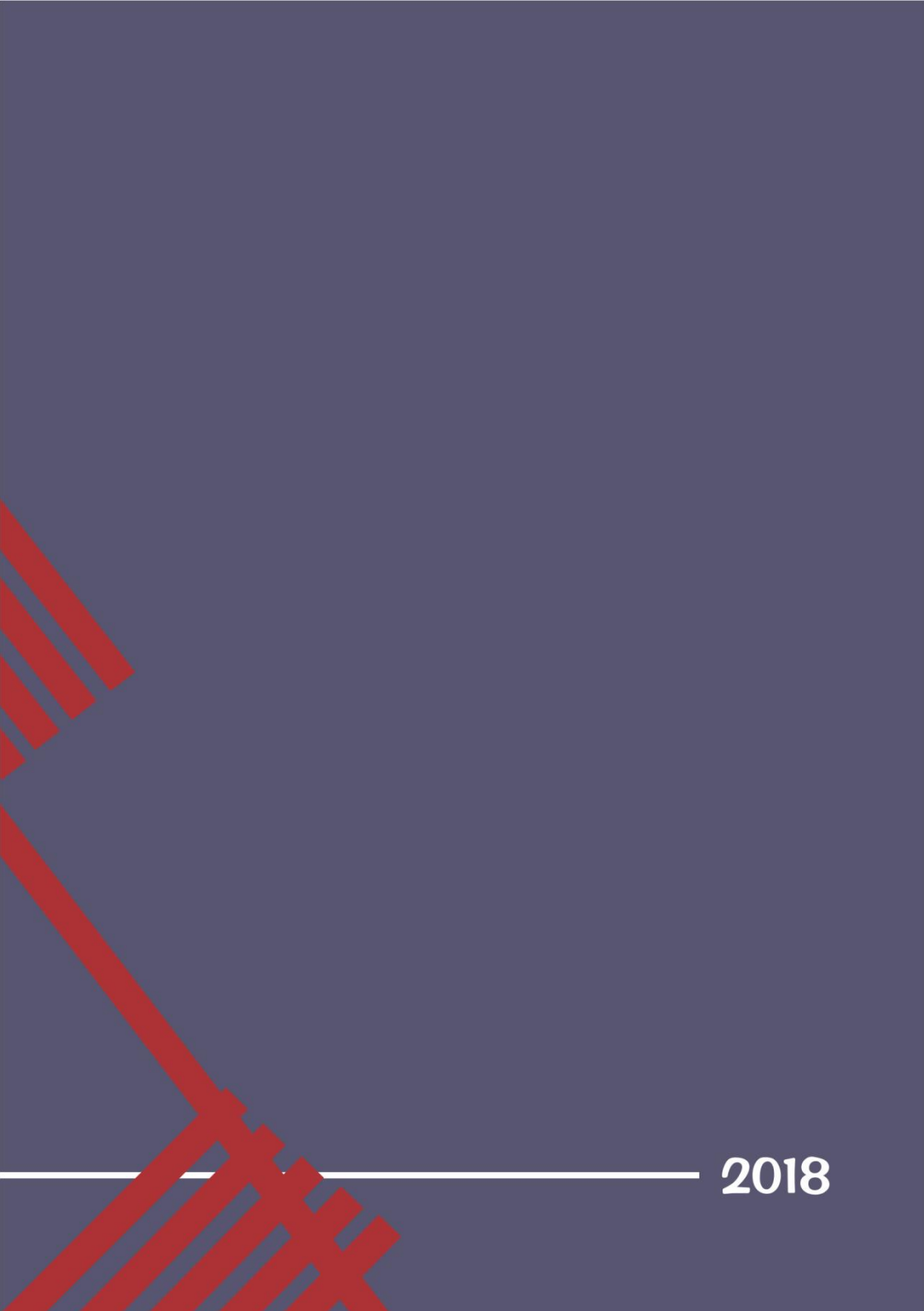
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