Introduction

The term congenital heart defects (CHD) include a variety of lesions with a wide spectrum of clinical importance, ranging from those of no functional or clinical significance to potentially life-threatening 'critical' lesions. **(Lloyd-Jones et al., 2009).**

Congenital heart defects are the most common group of congenital malformations, with a reported incidence of between 4 and 10 per thousand live-born infants, but a sizeable proportion is not detected by routine neonatal examination **(Wren et al., 2008).**

CHDs are one of the leading causes of infant death in the developed world, accounting for more deaths than any other type of malformation; up to 40% of all deaths caused by congenital malformation **(Lloyd-Jones et al., 2009) and** 6-10% of all infant deaths **(Rosamond et al., 2007).**

Most newborns with a CHD can be diagnosed using echocardiography and, if necessary, stabilized with prostaglandin infusion and treated with surgery or transcatheter intervention **(Mahle et al., 2009).**

Surgery has resulted in marked improvements in survival, particularly for those infants with potentially life-threatening conditions. However, if such defects are not detected sufficiently early then severe hypoxaemia, shock, acidosis and death are potential sequelae. Such cardiovascular compromise, if not lethal, can have significant long-term effects as a consequence of significant multiorgans insults, including hypoxic-ischemic brain injury. Timely recognition of these conditions is

likely to improve outcome and therefore the evaluation of screening strategies to enhance early detection is of great importance **(Mahle et al., 2009).**

Primary prevention is not possible; therefore, detection of CHDs prior to the onset of symptoms allows the possibility of interventions that may influence the natural history of the condition and subsequent outcome. As previously stated, this is of particular importance in infants with potentially life threatening, critical CHDs, most of whom are asymptomatic at birth**(Wren et al., 2008)**

Children with such life threatening defects may not initially have symptoms or the symptoms may be vague, and the condition is not detected on routine clinical examination in the majority of cases **(Mahle et al., 2009).**

The rationale for pulse oximetry screening is based on the fact that hypoxaemia is present, to some degree, in the majority of CHDs. This may result in obvious cyanosis; however, mild degrees of hypoxaemia cannot be detected by clinical observation, even by experienced clinicians The difficulty is exacerbated in infants with pigmented skin**(O'Donnell et al., 2007).**

Pulse oximetry was developed as a non-invasive method to determine arterial oxygen saturations (SpO2) and has been widely used in intensive care, operating theatres and emergency units. The ability to detect the different absorption spectra oxygenated and deoxygenated hemoglobin allows pulse oximeters to measure the amount of oxygen-saturated haemoglobin in the capillaries of an extremity, such as a finger or an ear lobe in an adult, or a hand or foot in a baby. Pulse oximetry thus

allows the detection of hypoxaemia that would necessarily produce invisible cyanosis; **(Mahle et al., 2008).**

Pulse oximetry has gained wide acceptance as a noninvasive method to determine oxygen saturation (SpO_2). The method does not require calibration and is able to provide data that correlate well with blood gas measurements **(O'Brien et al., 2005).**

Use of pulse oximetry has thus already contributed to heightened recognition of congenital heart disease in neonates **(O'Donnell et al., 2007).**

Aim of the work

This study was done on asymptomatic neonates to assess the value of pulse oximetry screening for detection of congenital heart disease and its incidence among asymptomatic neonates in Bab-EL-Sharia University Hospital.

Pulse Oximetry

Pulse oximetry was developed in the early 1970s based on the different absorption spectra between oxygenated and deoxygenated hemoglobin **(Aoyagi, 2003).** Deoxygenated hemoglobin absorbs light in the red band (600 to 750 nm), whereas oxygenated hemoglobin absorbs light in the infrared band (850 to 1000 nm). The ratio of light absorbance at these 2 wave lengths correlates with the saturation of hemoglobin in the capillaries **(Salyer, 2003).**

Pulse oximetry has gained wide acceptance as a noninvasive method to determine oxygen saturation (SpO_2). The method does not require calibration and is able to provide instantaneous data that correlate well with blood gas measurements **(O'Brien et al., 2000).**

Types of pulse oximetry:

Two types of pulse oximeter are available on the market. One measures functional Spo2 (oxyhaemoglobin / oxyhaemoglobin + reduced haemoglobin) and the other displays fractional Spo2 (oxyhaemoglobin / oxyhaemoglobin + reduced hemoglobin + approximated carboxyhaemoglobin and methaemoglobin). Functional Spo2 is thought to be about 1.6-2% higher at saturation levels used as cut offs in pulse imetry screening **(Poets and Southall, 1994).**

If this is not taken into consideration while screening, the actual cut off might shift by 1-2%, thus potentially increasing the false positive or false negative rates. However, the exact algorithms used for the calculation of fractional Spo2 are more complex, otherwise all fractional pulse oximeters would display a maximum of 98%, which is not the cause.Other technical differences between the various types of oximeter

include signal averaging times and methods for excluding movement artifacts' **(Poets and Southall, 1994).**

These are dependent on product developments aiming to minimize bias, which probably makes them less noteworthy. Adequate, up to date comparisons of the continually changing selection of oximeter brands, models and software upgrades on the market are not usually available. Despite the variations described above, the results of pulse oximetry screening seem rather consistent at cut off levels of about 94-96% allowing for minor technical differences**(Arlettaz et al., 2006).**

Uses of pulse oximetry:

Pulse oximetry is used routinely in the assessment of young children in neonatal intensive care units and emergency departments and has been proposed as an adjunct to the assessment of the newborn in the delivery room **(O'Donnell et al., 2007)** As such, some have proposed that pulse oximetry be considered as a vital sign equivalent in importance to pulse, respirations, and blood pressure **(Katzman, 1995).**

Pulse oximetry in neonatal period:

Beginning in the 1990s, investigators began to explore the possible role of neonatal oximetry in identifying CCHD that might otherwise go undetected. Initially, investigators demonstrated that in neonates with known CCHD, pulse oximetry measurements were significantly lower than in age-matched control subjects **(Hoke et al., 2002).**

Every baby is cyanotic until birth. The first breaths and increasing pulmonary circulation induce a rapid rise in pulse oximetry oxygen saturation (Spo2). The mean preductal (right hand) and postductal (foot)

Spo2 of healthy newborns at the age of 2 min has been shown to be 73% (range 44-95%) and 67% (34-93%), respectively **(Toth et al., 2002).** These values increased at 10 min to 92% (65-99%) and 89% (62-99%), respectively, and in every newborn both measurements reached 95% within 1 h. The reduction in the difference between preductal and postductal measurements reflects the diminishing ductal right to left shunt **(Toth et al., 2002).**

The normal baseline SpO_2 is quite stable at about 98%, except for short periods when it is lower, in particular during feeding and apnoeic spells **(O'Brien, 2000).**

Pulse oximetry in CHD:

Pulse oximetry is the first, appropriately simple method to have been used for universal screening of CHD, and the earliest reports (abstracts) on pulse oximetry screening were published in 1995 **(Kao et al.,1995).**

Hypoxemia is a common feature of many forms of congenital heart disease. It results from the mixing of systemic and venous circulations or parallel circulations as one might see in dextro-transposition of the great arteries. Hypoxemia may result in obvious cyanosis. However, generally, 4 to 5 g of deoxygenated hemoglobin is needed to produce visible central cyanosis, independent of hemoglobin concentration **(Lundsgaard and Van Slyke, 2002).**

For the typical newborn with a hemoglobin concentration of 20g/dL, cyanosis will only be visible when arterial oxygen saturation is <80%; if the infant only has a hemoglobin concentration of 10 g/dL, the saturation must be <60% before cyanosis is apparent **(Lees, 2003).** Importantly, those children with mild hypoxemia, with arterial oxygen

saturation of 80% to 95%, will not have visible cyanosis. Moreover, the identification of cyanosis is particularly problematic in black and Hispanic neonates because of skin pigmentation **(Lundsgaard and Van Slyke, 2002).**

Age at screening:

A slower disappearance of the shunt has been seen in 4-5% among larger populations, so the first hour of life is not suitable for pulse oximetry screening owing to the large number of false positive findings **(Richmond et al., 2002).** Thereafter, infants can be screened at any age, but somewhat more reliably after 2 h. First day screening identifies CHD and some other neonatal problems, whereas later screening is more specific for CHD **(Meberg, 2005).**

Probe site:

Newborns that have obligatory or mixing cyanotic CHD have reduced SpO_2 both preductally and postductally. Defects with ductal dependent systemic circulation exhibit higher preductal than postductal SpO_2 due to right to left shunting of unoxygenated blood to the lower body Pulse oximetry screening is thus most effective with postductal probe placement **(Hoke et al., 2002).**

Signal quality and newborn behavior:

Technical errors are minimized by accepting only high quality pulse oximetry signals confirmed by a good pulse signal displayed simultaneously **(Arlettaz et al., 2006).** Measurements should not be made when the infant is moving, crying, eating or has an apnoeic spell **(O'Brien, 2000).** The heart rate displayed by the oximeter should be within the range expected for a calm, regularly breathing newborn (about

90-160/min). The highest stable SpO_2 value, maintained longer than the averaging time of the pulse oximetry, is recorded. In practice, the measurement usually takes a few minutes **(Bakr and Habib, 2005).**

The Transition From Fetal to Neonatal Circulation

What is the neonatal period?

Fetal and extrauterine life form a continuum during which human growth and development are affected by genetic, environmental, and social factors. The perinatal period is most often defined as the period from the 28^{th} wk of gestation through the 7^{th} day after birth (additional definition include the 20^{th} wk of gestation to the 7^{th} day and the 20^{th} wk of gestation to the 28^{th} day). The neonatal period is defined as less than 28days of life and may be further subdivided into:

Period 1: birth to less than 24hr,
Period 2: 24 hr to less than 7 days,
And period 3: 7 days to less than 28 days

(Carlo, 2011).

What is the fetal circulation ?

The fetal circulation consists of parallel pulmonary and systemic pathways in contrast to the circuit of the normal postnatal circulation. Oxygenated blood returns from the placenta through the umbilical vein and enters the portal venous system. A variable amount of this stream bypasses the hepatic microcirculation and enters the inferior vena cava by way of the ductus venosus. Inferior vena caval blood is from the ductus venosus, hepatic veins, and lower body venous drainage and is partly deflected across the foramen ovale (FO) into the left atrium **(Webb et al., 2005).**

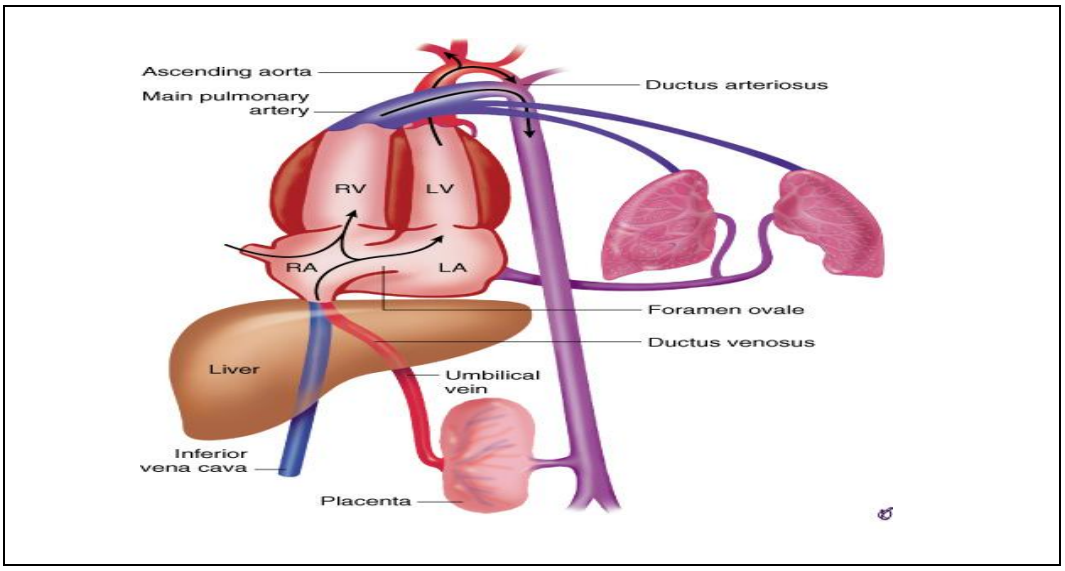

Fig. (1): The fetal circulation **(Webb et al., 2005).**

Almost all superior vena caval blood passes directly through the tricuspid valve, entering the right ventricle. Most of the blood that reaches the right ventricle bypasses through the ductus arteriosus into the descending aorta.. The major portion of blood ejected from the left ventricle supplies the brain and upper body, with lesser flow to the coronary arteries; the balance passes across the aortic isthmus to the descending aorta, where it joins with the large stream from the ductus arteriosus before flowing to the lower body and back to the placenta **(Smallhorn et al., 2005).**

Fetal pulmonary circulation:

In fetal life, the alveoli are fluid filled, and the pulmonary arteries and arterioles have relatively thick walls and a small lumen, similar to arteries in the systemic circulation. The low pulmonary blood flow in the fetus (7 to 10 percent of the total cardiac output) is the result of high pulmonary vascular resistance. Fetal pulmonary vessels are highly reactive to changes in oxygen tension or in the pH of blood perfusing

them as well as to a number of other physiological and pharmacological influences **(Webb et al., 2005).**

In the late gestation, pressures in the fetal left and right ventricles are the same (approximately 70 mmHg systolic). The pressure is transmitted into the pulmonary arteries and ascending and descending aorta. The left and right sides of the fetal circulation function more or less parallel, that is a major distinction from the postnatal (in series) circulation. Combined ventricular output that passes through the placenta is more or less equal to the volume of right ventricular blood that passes through the lungs after birth **(Rudolph, 2008).**

Transitional circulatory compromise:

The transitional circulatory changes in the first 12 to 24 hours after birth are a period of unique circulatory vulnerability for the extremely preterm infant. During normal postnatal adaptation, pulmonary vascular resistance falls, systemic vascular resistance rises when the placenta is removed from the circulation, the ductus arteriosus closes, and the FO is closed by the reversal of the arterial pressure gradient. As these changes occur, the left ventricle has to double its output. Given that very preterm infants' cardiovascular systems are adapted to the low-resistance intrauterine environment and that their myocardium is immature, it is not surprising that these babies often have difficulties during this critical period **(Evans and Seri, 2005).**

Furthermore, blood pressure in the low-normal to normal range does not necessarily translate into normal organ blood flow and tissue perfusion in these patients. Indeed, it is only when mean blood pressure is higher than 40mmHg during the first postnatal day in the preterm neonate born before 30 weeks of gestation that the systemic blood flow can

definitely be assumed to be normal **(Osborn et al., 2004)**.

Another special characteristic of the cardiovascular adaptation in this patient population is that early shunts through the preterm ductus arteriosus and FO are not balanced and, thus, can produce left-to-right shunts of significant clinical importance. The immediate postnatal physical constriction of the ductus is characterized by great variation, but if constriction fails, very large shunts can occur within a few hours of birth, leading to high, not low, pulmonary blood flow as previously thought. These abnormal hemodynamic changes may be further augmented by the administration of surfactant **(Evans and Seri, 2005).**

Congenital Heart Diseases

Definition of CHD:

The term congenital heart defects (CHD) encompass a variety of lesions with a wide spectrum of clinical importance, ranging from those of no functional or clinical significance to potentially life-threatening 'critical' lesions. If undiagnosed, infants with a critical lesion are at risk of acute cardiovascular collapse or death **(Lloyd-Jones et al., 2009).**

Epidemiology of CHD:

Congenital heart defects are the most common group of congenital malformations**(Wren et al., 2008).** About 0.5 - 0.8 % of all children are born with congenital heart diseases (about 8-10 out of every 1000 children every year) **(Fixler et al., 2009).**

Congenital heart diseases have a wide spectrum of severity of symptoms in infants (about 2-3 in 1000 newborn infants will be symptomatic with heart diseases in the first year of life) **(Nakazawa et al., 2005).**

The diagnosis is established by the first week of age in 40-50% of patients with congenital heart diseases and by the first month of age in 50-60% of patients **(Moller and Hoffman, 2005).**

Ventricular septal defect is generally considered to be the most common type of malformations accounting for about 1/3 of all congenital heart defects followed by patent ductus arteriosus and bicuspid aortic valve **(Campbell, 2008).**

More than forty thousand babies allover the world are born each year with a congenital heart disease; 4000 will survive their first year (the children's heart foundation) **(Ferencz et al., 2008).**

Twice as many children die each year from a congenital heart disease than all forms of pediatric cancers (the children's heart foundation) **(Mirchell et al., 2006).**

With advances in both palliative and corrective heart surgery, the number of children with congenital heart diseases surviving to adulthood has increased dramatically. Despite theses advances, congenital heart diseases remain the leading cause of death in children with congenital malformations **(Correa-Villansenor et al., 2008).**

Table (1) Relative frequency of major congenital heart lesions:

lesion	***% of all lesions***
Ventricular septal defect	30-35
Atrial septal defect	6-8
Patent ductus arteriosus	6-8
Coarctation of aorta	5-7
Tetralogy of Fallot	5-7
Pulmonary valve stenosis	5-7
Aortic valve stenosis	4-7
D-transposition of great arteries	3-5
Hypoplastic left ventricle	1-3
Hypoplastic right ventricle	1-3
Truncus arteriosus	1-2
Total anomalous pulmonary venous return	1-2
Tricuspid atresia	1-2
Single ventricle	1-2
Double outlet right ventricle	1-2
Others	5-10

Excluding patent ductus arteriosus in preterm neonates, bicuspid aortic valve, physiological peripheral pulmonary stenosis and mitral valve prolapse **(Ferencz et al., 2008).**