The Cardiothoracic Surgeon

REVIEW

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The management of newborns with critical congenital heart diseases prior to transport to a cardiac center

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Abstract

Critical congenital heart diseases (CCHD) are important causes of mortality and morbidity in the newborn period. Even after diagnosis, their management could be seriously compromised by the unplanned delivery in hospitals with limited expertise and resources. The newborn may spend days or week before transport putting a significant burden on the neonatal team to manage such challenging diseases. In this review, the management principles of each individual pathology are discussed in the setting before transport to cardiac centers. Understanding these principles will help the treating teams evaluate and manage those complex conditions. The review avoids the advanced discussion on the management of CCHDs not applicable to the pre-transport setup. It highlights the critical elements in the maintenance of normal hemodynamics specific to each pathology and their variation.

Keywords Critical congenital heart, Cyanosis, Newborns, Congenital, Hemodynamics, Critical

Background

Congenital heart diseases continue to be the most common congenital anomalies in newborn children with an incidence of 8 in 1000 live births [1]. Their impact on the newborn health varies from being simple to life-threatening. Understanding the hemodynamic principles of critical congenital heart diseases (CCHD) is vital for their management to prevent unfortunate mortality or morbidity [2]. This task is not simple, and it takes dedicated learning and experience to master various anomalies with heterogenic combinations of critical pathologies. The challenge is further compounded by the inability to deliver those pregnancies in centers where the expertise is available to manage such conditions due to limitations in fetal echocardiography [3]. Moreover, the transfer of newborn with CCHD is challenging in many countries with limited resources and bed availability [4, 5]. Newborns with CCHD may spend several days to weeks in a neonatal intensive care unit (NICU) with limited support from pediatric cardiology and cardiac surgery services. Fortunately, most CCHD can be kept a live with no additional morbidities for weeks with appropriate use of Prostaglandin E1 (PGE1) and respiratory support that can be provided in almost all neonatal care units. In this article, we will review the important management strategies for CCHD in the NICU before the baby is transferred to a cardiac center.

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Main text

General considerations

The surveillance of newborn with CCHD starts by reviewing the maternal medical history for risk factors for congenital heart diseases and reviewing the fetal ultrasound or fetal echocardiogram. Physical examination is

paramount in the initial admission to the nursery and prior to the discharge with emphasis on cyanosis, tachypnea, murmur, and palpable femoral pulse[6]. The physical examination of newborn with CCHD may not reveal clear abnormality due to transition from fetal circulation with elevated pulmonary vascular resistance (PVR) and the presence of the patent ductus arteriosus (PDA). The newborn pulse oximetry screening for CCHD should be performed in an optimal condition and interpreted carefully [7]. When there is any doubt on the presence of CCHD the pediatric cardiologist should be consulted before discharging the newborn. Newborns who appear to be normal in the nursery may present in very critical conditions in two weeks with potentially long term sequalae.

Although prostaglandin E1 (PGE1) infusion is the mainstay intervention in the majority of CCHD conditions it does not help or may not be enough in some of these conditions. In other conditions where the circulations are balanced, using PGE1 is potentially harmful and will prolong the hospitalization of the potentially dischargeable and stable newborn. Similarly, oxygen therapy may not be needed in a balanced circulation with mixing lesions. Lower target oxygen saturation that is appropriate for the physiology is the ideal approach. Blind oxygen therapy to achieve saturation above 90% in an excessive pulmonary blood flow (PBF) physiology will unnecessarily prolong the respiratory support and the hospital stay. Lastly, unless there is documented ventricular dysfunction by echocardiogram, inotropic infusions should not be utilized for the sole purpose of improving the cardiac function especially if the blood pressure is acceptable. Inappropriate overuse of inotropic infusions changes the circulation balance and compromise the systemic blood flow with potential serious consequences on the gut and kidneys perfusion[8, 9].

Left to right shunt

Large ventricular septal defect (VSD), single ventricle, aorto-pulmonary window, and large patent ductus arteriosus (PDA) are the most common causes of significant left to right shunt in the newborn. They cause significant pulmonary congestion and systemic hypoperfusion starting few days after birth in many cases. Classically, they have normal oxygen saturation due to excessive pulmonary blood flow (PBF); however, they may have lower oxygen saturation due to elevated pulmonary vascular resistance or pulmonary edema. Oxygen is a potent pulmonary vasodilator and should be avoided in such cases unless there is significant desaturation. Acceptable saturation of 88% may be needed to avoid using excessive oxygen and allow for discharge from the NICU. Diuretic therapy, high caloric feeding, and fluid restrictions are the main stay of management [10, 11]. The newborn is expected to have some residual tachypnea and slow weight gain that can be followed and managed in the outpatient clinic. In few cases, the newborn cannot be discharged and will need to be transferred for surgical or catheter intervention.

Total anomalous pulmonary venous drainage

Total anomalous pulmonary venous drainage (TAPVD) is usually not obstructed in the cardiac type where the pulmonary venous confluence (PVC) is connecting to the coronary sinus and almost always obstructed in the infracardiac type where the PVC connects via vertical vein to the systemic veins below the diaphragm. In the supracardiac type where the PVC connects via vertical vein to the innominate vein and mixed type the presence of obstruction needs to be carefully evaluated by echocardiogram. In the presence of obstruction, the newborn will develop progressive pulmonary venous edema and pulmonary hypertension. The veins need to be repaired surgically upon discovery of the obstruction [12, 13]. Because the pulmonary blood flow through the pulmonary arteries is normal, the ductus arteriosus is not needed and prostaglandin may increase the pulmonary congestion. In the rare instances where the obstruction is at the level of the ductus venosus prostaglandin may improve the obstruction; however, it should not be regarded as a standard short-term therapy as the standard management for obstructed TAPVD is surgical repair. This is one of the very few pathologies where most of the temporarily stabilizing measures applied in other conditions will not work and the newborn should be transferred immediately to a facility where surgery can be done. In the case of unobstructed cardiac or supracardiac types, the newborn can be discharged home safely with acceptable saturation of 75% or higher and follow up with pediatric cardiology in 2–4 weeks.

Critical pulmonary valve stenosis and pulmonary atresia

In critical pulmonary stenosis, the newborn cannot maintain acceptable saturation without the PDA. In both conditions, PGE1 infusion must be maintained till a more stable source of PBF is established via pulmonary valve dilation/perforation, PDA stenting, or BT shunt [14]. PGE should be started as soon as the diagnosis is suspected at dose 0.03 to 0.05 μm/kg/min in separate intravenous line from other infusions and should be evaluated regularly for patency and adequate flow. The dose can be increased to 0.1 μm/kg/min if the saturation of 75% can not be achieved. Care should be taken to observe for apnea that may require mechanical respiratory support. In rare occasions when the PDA could not be opened,

we used bolus PGE1 3 to 5 μm/kg over 10 min in a small number of cases with success. PGE1 infusion could lower the blood pressure, and the newborn may need inotropic support with either dopamine or epinephrine. The newborn may develop pulmonary over circulation due to large PDA suggested by congested lung on chest X-ray and/or development of metabolic acidosis. In such case, the PGE1 should be decreased to the lowest possible dose of 0.01 to 0.005 μm/kg/min as long as the saturation is maintained above 75%. Upon transport of such cases care must be taken not to interrupt the PGE1 flow as this may have fatal consequences in an already limited setup.

Tricuspid atresia

In tricuspid atresia (TA), there is obligatory right to left shunt at the atrial level. Rarely, the atrial communication is restrictive and the newborn will need balloon atrial septostomy. The connection of the great arteries and the presence of pulmonary stenosis are the determinants of the newborn hemodynamics [15]. If there is pulmonary atresia or critical pulmonary stenosis, the PDA must be kept open to provide for pulmonary blood flow. If the aorta is arising from the right ventricle (RV), the systemic flow is dependent on the size of the VSD. The VSD in TA is usually large but may become restrictive with time in the first few months. If the aorta is arising from the LV and there is no pulmonary stenosis, the physiology is similar to large VSD with the left to right shunt beyond the first week or two. In the presence of pulmonary stenosis, it is recommended to keep the newborn admitted till the PDA is closed or become significantly small and observe the saturation. If the saturation is above 75%, the baby can be discharged safely. In TA with malposed great arteries and adequate size VSD for systemic flow and in TA with normally related great arteries with no pulmonary stenosis, the newborn can be discharged from the nursery after 24 to 48 h of routine observation.

Tetralogy of Fallot

Tetralogy of Fallot (TOF) in newborn is a spectrum of limited pulmonary blood flow (PBF); from normal flow with no or little obstruction to total atresia [16]. It is an abnormality of the branch pulmonary arteries as well as the heart with various degrees of hypoplasia. The newborn management depends on the degree of pulmonary stenosis and the source of pulmonary blood flow in case of pulmonary atresia. If there is mild to moderate pulmonary stenosis, the newborn can be discharged with follow up in 2–4 weeks to re-evaluate the degree of pulmonary stenosis after the pulmonary vascular resistance has decreased. If the pulmonary stenosis is severe, the newborn should be kept in hospital and saturation observed until the PDA is closed. If saturation is maintained at or above 75%, the newborn can be discharged otherwise prostaglandin E1 should be started in preparation for more reliable source of PBF with Blalock-Taussing shunt (BT shunt), PDA stent, right ventricular outflow (RVOT) stent, or neonatal repair [17–19]. There should be no delay in starting prostaglandin if saturation is not adequate because the longer the delay the less likely the PDA will respond to the treatment. Other measures of supporting the circulation while waiting for emergency BT shunt are fluid boluses and inotropes to elevate the systemic vascular resistance and augment PBF. In this context, diuretic therapy should be avoided as it reduces the preload needed to overcome the RVOT obstruction. In pulmonary atresia or if one of the branch pulmonary arteries is discontinuous and supplied by a PDA, needless to stress, the importance of prostaglandin E1 till a reliable BPF is established to both lungs [20]. In case of pulmonary atresia and major aortopulmonary collateral to both lungs, the newborn can be discharged safely if saturation is more than 75%. Time and effort should be taken to carefully exclude the possibility of the PDA supplying one of the branch pulmonary arteries as this will lead to loss of that branch if the PDA closes [21].

TOF with absent pulmonary valve is a rare condition where the valve annulus is small, but the leaflets did not form causing significant stenosis and regurgitation. By the end of the pregnancy, the fetus will have huge pulmonary arteries causing significant bronchomalacia and compression on the airways. If they are born with severe airway obstruction, their management is challenging and the prognosis is not favorable [22]. They should undergo CT angiography to evaluate the size of the pulmonary artery and the degree of bronchial obstruction. Their discharge vs transport decision depends on the degree of the respiratory distress caused by the airway obstruction.

Ebstein anomaly

In mild or moderate Ebstein anomaly of the tricuspid valve, the newborn will likely be asymptomatic; however, in severe Ebstein, the right atrial (RA) pressure is elevated and may cause systemic congestion with edema, ascites, and hepatomegaly [23, 24]. The abnormal valve may cause functional pulmonary atresia necessitating the use of prostaglandin to maintain adequate saturation of $\geq$ 75%. Diuretic therapy is needed to improve the edema but should be carefully used as the pulmonary flow may be load dependent.

Double outlet right ventricle

Double outlet right ventricle (DORV) physiology is similar to TOF physiology if there is severe pulmonary stenosis and similar to large VSD physiology if the VSD is subaortic without pulmonary stenosis both discussed

above [25]. A third entity is similar to transposition of the great artery physiology if the VSD is subpulmonary (Taussing-Bing Anomaly) and will be discussed in the following sections. The TGA type could be associated with posterior deviation of the conal septum causing left ventricular outflow tract obstruction and commonly associated with hypoplastic aortic arch or coarctation of the aorta.

Transposition of the great arteries

Transposition of the great arteries (TGA) has very good long-term outcome after successful arterial switch surgery in the current time. Unfortunately, it has one of the highest mortalities in the newborn period if the diagnosis is not established promptly or the hemodynamic inadequacies are not addressed [26, 27]. Newborn with TGA and large atrial septal defect (ASD) usually have acceptable saturation of more than 75% and can have their surgery in the first few weeks of life. It is wise to keep them on PGE until they are transported to a facility where expertise in managing such cases is available. Newborn with TGA who do not saturate well after starting PGE are either having restrictive patent foramen ovale (PFO) or have pulmonary hypertension (PHTN). Balloon atrial septostomy is the ideal procedure to establish mixing in the atrial level if the PFO is small and should be performed, if possible, in the peripheral hospital or the baby transported in the first few hours after establishing the diagnosis. Those with desaturation despite acceptable size ASD and an open PDA are likely having PHTN [28]. They need to be managed with adequate sedation, paralysis, and mechanical ventilation to achieve normal carbon dioxide (CO_2) level in the arterial blood (35 to 45 mmHg) and normal bicarbonate (HCO_3). They need to be started on nitic oxide (NO) and a maximum oxygen concentration. Milrinone should be utilized for its pulmonary vasodilator effect and other inotropes (dopamine or epinephrine) should be titrated to achieve normal blood pressure. The earlier these measured are used in combination with careful attention to avoid high CO_2 and metabolic acidosis the more likely the baby will survive with no major sequalae. These measures need to be continued till the baby is stable enough for surgery and may be needed for few days after the surgery. Newborns with large VSD are likely to have adequate mixing; however, this need to be verified before discharge by observing the baby's saturation until the PDA is closed.

Critical aortic stenosis

In critical aortic stenosis (AS), the flow through the aortic valve is inadequate to support the systemic circulation. They have very high LV pressure and low aortic pressure causing serious coronary hypoperfusion. The newborn will need the PDA to maintain perfusion and blood pressure. The LV obstruction causes variable degrees of hypertrophy, dilation, dysfunction, or hypoplasia of the left ventricle. The newborn is at risk of ventricular arrhythmia which add to the serious risk of theses newborns. The lung is congested due to elevated left atrial (LA) pressure and will get worse with time if the obstruction is not relieved. Prostaglandin infusion to keep the PDA open is essential but it does not improve the elevated LA pressure. The associated pulmonary hypertension should never be treated with pulmonary vasodilators like nitric oxide as this will cause increased flow to an already obstructed venous drainage. Adequate size ASD will improve the pulmonary edema; however, atrial septostomy is not a standard procedure in this condition as the same effect can be achieved by aortic valve balloon dilation. The newborn may need small dose of inotrope to augment the blood pressure and oppose the sedation effect.

In severe aortic stenosis, milder degree of the same hemodynamic usually manifests, but the baby does not need the PDA to maintain systemic perfusion. They are at risk of pulmonary edema, ventricular dysfunction, and arrhythmia caused by coronary hypoperfusion. After a period of observation, some of them may be stable enough to be discharged and evaluated in a cardiac center in 1–2 weeks with balloon dilation as soon as possible in the first month of life. Careful assessment in collaboration with the accepting center should be considered before discharging such newborn [29].

Hypoplastic left heart syndrome

The hemodynamics of hypoplastic left heart syndrome (HLHS) is the most challenging of all CCHD [30]. The systemic circulation is dependent on the PDA. The ratio of blood flow to the pulmonary and systemic circulation (Qp:Qs) is dependent on the pulmonary vascular resistance which decreases over days to weeks accompanied by increase in the PBF and systemic hypoperfusion. Oxygen saturation of 75 to 80 % is simple but crude measure of the relatively balanced flows. A higher saturation means low systemic perfusion and should be avoided. The lower oxygen saturation towards 75 to 80%, the more balanced are the Qp:Qs and oxygen therapy should be avoided if these targets are met. If the atrial communication is small, the newborn will develop venous pulmonary congestion but will have less pulmonary over circulation. These changes are dynamic and are consistently changing by factors that manipulate pulmonary and systemic resistance like oxygen, CO_2, PH, mechanical ventilation, inotropes, and temperature. Most newborn will develop progressively increasing need for diuretics due to lung congestion. Because the perfusion of the gastrointestinal

tract and kidney could be compromised feeding should be carefully monitored and effort should be made to avoid kidney injury. Trophic feeding is shown to be advantageous before surgery [31, 32]. The ethics of do not resuscitate decision (DNR) is an important consideration in this condition before and after birth and should be discussed with the treating cardiologist [33]. The outcome of Norwood surgery for this condition is less favorable with prematurity, genetic syndrome, severe tricuspid valve regurgitation, and right ventricular dysfunction [34–36]. These factors should be considered before committing the family to a transfer to another city or province for potentially prolonged hospital stay. If Norwood is considered, a serious effort need to be made to transport the newborn as early as possible because delaying Norwood surgery beyond three weeks increases the risk of operative mortality and significant morbidity [37].

Coarctation of aorta and interrupted aortic arch

In severe coarctation (COA) or interrupted arch (IAA), the blood supply to the body parts that are supplied by branches distal to the COA or IAA is dependent on the PDA. Classically, the lower body parts have lower saturation, however with adequate perfusion if the PDA size is large enough. If discovered early, the newborn will have normal function and does not need more than prostaglandin infusion. Newborns who present at later stage may develop severe left ventricular (LV) dysfunction and need more support with ventilation and inotropes.

Table 1 summarizes the various dominant physiological states of CCHD and the most important considerations for their management in the peripheral hospitals.

Combination of lesions

Unfortunately, many newborns have combination of more than one anomaly that necessitate careful understanding of the relative severity of each one and its relative weight on affecting the overall hemodynamics. As a guiding role, significant obstruction in the systemic circulation starting from the pulmonary veins to the descending aorta should be addressed before discharge. Inadequate pulmonary blood flow is reflected on the saturation, and PGE1 should be continued if saturation is less than 75%. On the contrary, in the case of mixing lesions, if the pulmonary flow is not limited by significant obstruction, the newborn will develop significant

Table 1 Summary of the dominant hemodynamic physiologies of CCHD and their management

Dominant physiology	Pathology	Management
Left to right shunt	AVSD, VSD, PDA, AP window	Diuretic therapy Accept saturation > 88% (usually >92%) Avoid unnecessary oxygen therapy Fluid restriction Avoid hyperventilation and alkalosis
Obstructed pulmonary veins	Obstructed TAPVR	Immediate transport for cardiac surgery
Cyanosis with unlimited PBF	Unobstructed TAPVR, Tricuspid atresia with no PS, Truncus arteriosus, single ventricle, DORV with no PS	Diuretic therapy Accept saturation > 75% Discharge after ensuring there is no significant PS and the PDA is almost closed Early follow-up in cardiac center
Cyanosis with limited BPF	Pulmonary atresia, severe PS, TOF, DORV with severe PS, Tricuspid atresia with severe PS, severe Ebstein anomaly	Prostaglandin E1 infusion Accept saturation > 75% Increase the intravascular volume Vasopressors to augment pulmonary flow Transport to a cardiac center
Parallel circulations	TGA with intact ventricular septum, DORV with malposed great arteries	Prostaglandin E1 infusion Accept saturation > 75% Evaluate the need for balloon atrial septostomy Treat pulmonary HTN aggressively if desaturated with adequate ASD. Transport to a cardiac center
Obstructed systemic circulation	HLHS, Tricuspid atresia with malposed great arteries and small VSD, Aortic stenosis, Interrupted aortic arch, Coarctation of the aorta	Prostaglandin E1 infusion Accept saturation > 75% Avoid hyperventilation, oxygen therapy and alkalosis. Fluid restriction Utilize ventilation with higher PEEP Transport to a cardiac center

AVSD Atrioventricular septal defect, *VSD* Ventricular septal defect, *PDA* Patent ductus arteriosus, *AP window* Aorto-pulmonary window, *TAPVR* Total anomalous pulmonary venous return, *PBF* Pulmonary blood flow, *PS* Pulmonary stenosis, *DORV* Double outlet right ventricle, *TOF* Tetralogy of Fallot, *TGA* Transposition of the great arteries, *HLHS* Hypoplastic left heart syndrome, *PEEP* Positive end expiratory pressure

pulmonary over circulation and systemic hypoperfusion. Oxygen saturation of ≥75% should be adequate in mixing lesions and additional oxygen therapy will likely be harmful as they cause pulmonary over circulation and systemic hypoperfusion.

Finally newborn with CCHD are at risk for other anomalies in the airways, lungs, skeleton, brain, spinal cord, gastrointestinal tract, and urinary system. Carful assessment of these anomalies and their contribution to the clinical or hemodynamic condition is paramount before assuming that the heart is the main cause of the clinical or hemodynamic state.

Conclusions

The health and survival of newborns with CCHD are vulnerable in the period before the diagnosis is established and during their hospital stay in the facility not equipped to manage their conditions. Until the transport can be done in a matter of hours, neonatologists and pediatricians need to master the essential principles of managing these conditions in the pre-transport setup. Although the understanding of hemodynamics in various CCHD is difficult task and need trained professional, the practical concepts in their management before surgery are simple and can be mastered by neonatologist and pediatrician with minimal effort. The pre-transport period of newborn with CCHD is a serious vulnerability in the health system in the Kingdome of Saudi Arabia and many developing countries, and additional efforts should be made to understand it and overcome its challenges.

Abbreviations

CCHD	Critical congenital heart diseases
NICU	Neonatal intensive care unit
PGE1	Prostaglandin E1
AVSD	Atrioventricular septal defect
VSD	Ventricular septal defect
PDA	Patent ductus arteriosus
PBF	Pulmonary blood flow
TAPVD	Total anomalous pulmonary venous drainage
PS	Pulmonary stenosis
PVC	Pulmonary venous confluence
TA	Tricuspid atresia
TOF	Tetralogy of Fallot
BT shunt	Blalock-Taussing shunt
RVOT	Right ventricular outflow tract
CT	Computerized tomography
RA	Right atrium
DORV	Double outlet right ventricle
TGA	Transposition of the great arteries
ASD	Atrial septal defect
PHTN	Pulmonary hypertension
PFO	Patent foramen ovale
AS	Aortic stenosis
LA	Left atrium
DNR	Do not resuscitate
COA	Coarctation of the aorta
IAA	Interrupted aortic arc

Acknowledgements
None.

Authors' contributions
Dr. Naif Alkhushi is the only author. The author read and approved the final manuscript.

Funding
No funding was provided for this review.

Availability of data and materials
Not applicable for the article.

Declarations

Ethics approval and consent to participate
Not applicable for a review article.

Consent for publication
No patient data or information submitted.

Competing interests
The author declares that they have no competing interests.

Received: 5 November 2022 Accepted: 20 December 2022
Published online: 05 January 2023

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