January: Question 10

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You are asked to evaluate a newborn with a heart murmur. An echocardiogram reveals that the infant has normal cardiac anatomy with a persistent left superior vena cava (LSVC).

Of the following, the MOST likely structure into which a LSVC drains directly is the:

1	coronary sinus
2	left atrium
3	left brachiocephalic vein
4	right atrium
5	right superior vena cava

You selected 3, the correct answer is 1.

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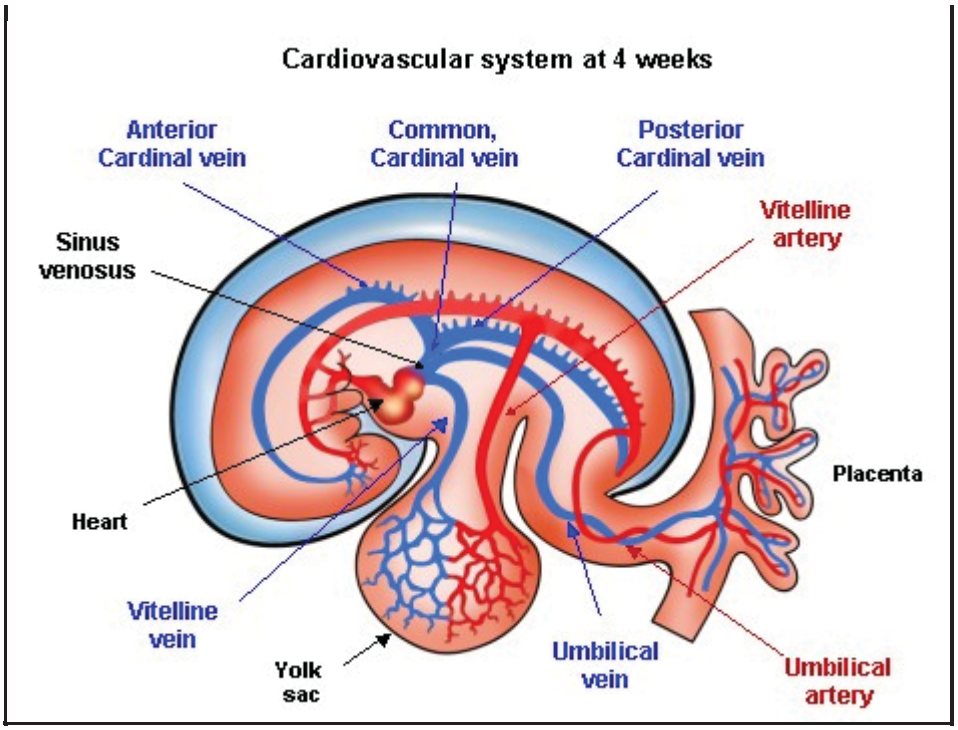
The embryologic development of the heart and great vessels is a complex process. The primordial heart and vascular system first appear in week 3 of embryonic development, and the heart begins to beat at 22 to 23 days. The vascular system develops in a bilateral symmetric fashion and undergoes a sequence of obliterations, remodeling, and anastomoses. Obliterations tend to occur on the left side of the embryonic venous system and the right side of the arterial system. Obliterations of embryonic left-sided venous structures results in venous drainage being channeled to the right atrium. Vascular malformations can arise at many stages of development. Failure of obliteration of left-sided venous structures results in a persistent left superior vena cava (LSVC).

Three major sets of paired veins drain into the tubular heart of a 4-week embryo (Figure 1).

Figure 1:Cardiovascular system at 4 weeks.

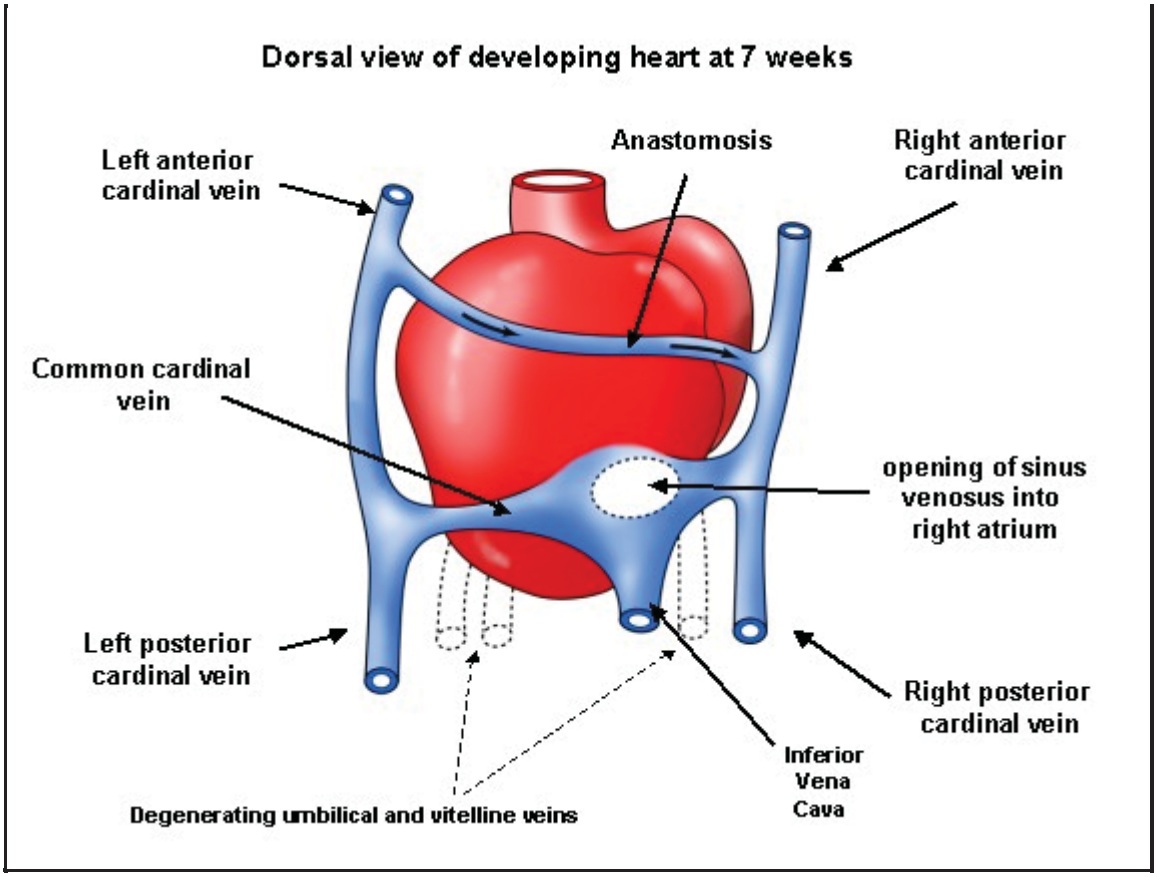


The left and right umbilical veins carry oxygenated blood from the primordial placenta; the left and right vitelline veins return poorly oxygenated blood from the yolk sac; and the left and right cardinal veins return poorly oxygenated blood from the body of the embryo.

The left and right cardinal veins, the main venous drainage system of the embryo, are further subdivided into anterior cardinal veins that drain the cephalic part of the embryo, and posterior cardinal veins that drain the caudal part of the embryo. The left and right anterior and posterior cardinal veins join as the common cardinal veins and drain into the sinus venosus (coronary sinus) which in turn drains into the primordial atrium.

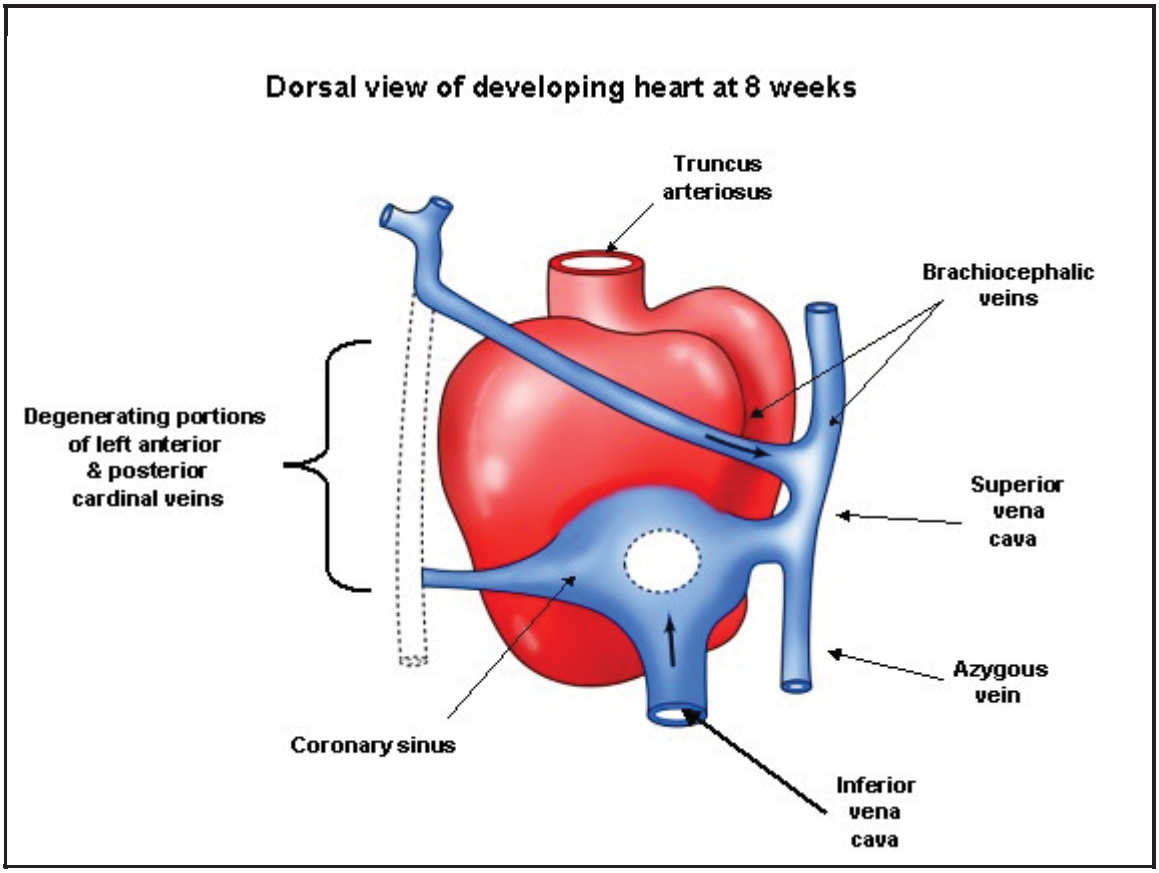
At week 7 of gestation, the left and right anterior cardinal veins are connected by an oblique anastomosis that shunts blood from left to right (Figure 2).

Figure 2:Dorsal view of developing heart at 7 weeks.



At week 8 of gestation, the caudal portion of the left anterior cardinal vein degenerates (Figure 3).

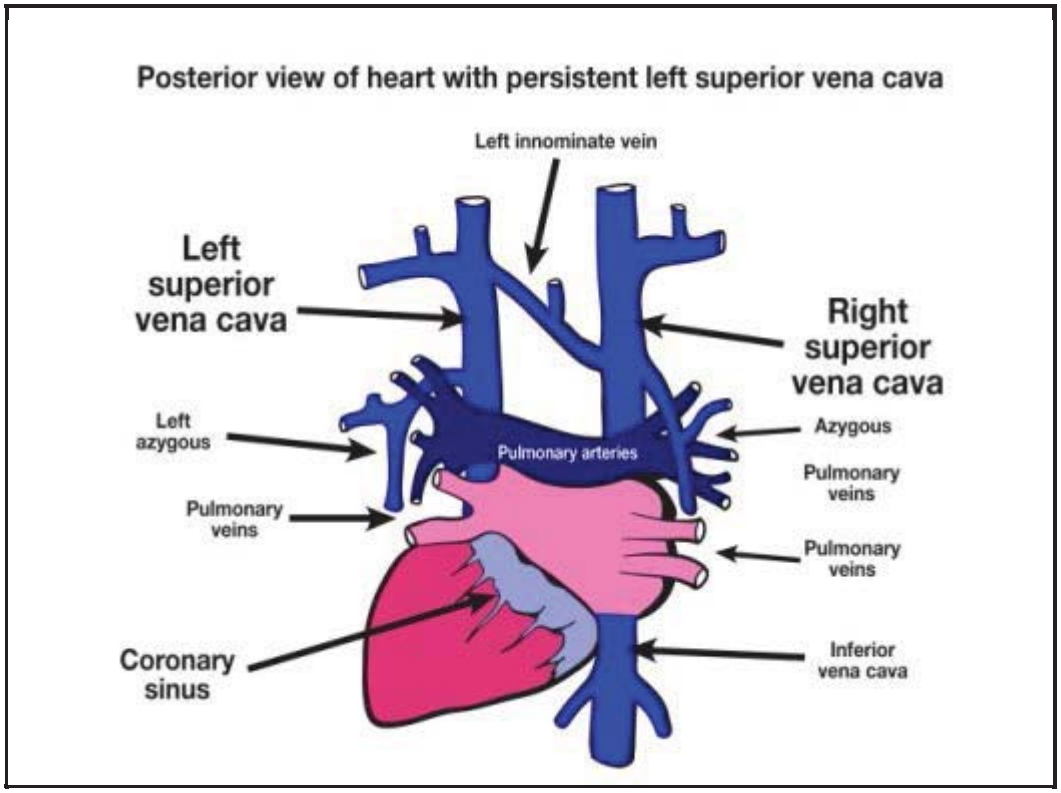
Figure 3: Dorsal view of developing heart at 8 weeks.



The cephalic portion of the left anterior cardinal vein and the oblique anastomotic shunt become the left brachiocephalic vein. The right anterior cardinal vein and right common cardinal vein become the superior vena cava. Failure of obliteration of the caudal portion of the left anterior cardinal vein results in a persistent LSVC.

A persistent LSVC is the most common malformation of systemic venous drainage. A persistent LSVC is present in 0.4% of the population and 10% of patients with congenital heart disease. The LSVC starts at the junction of the left internal jugular and left subclavian vein, passes lateral to the aortic arch, and receives blood from the left superior intercostal vein and hemiazygous system. In approximately 90% of cases, the LSVC drains into the coronary sinus which in turn drains into the right atrium (Figure 4).

Figure 4: Posterior view of heart with persistent left superior vena cava.



Although drainage of a LSVC into the coronary sinus causes dilation of the coronary sinus, it is usually asymptomatic. The adverse events that have been reported include cardiac arrhythmias due to stretching of the atrioventricular node, and obstruction of left ventricular flow because of partial occlusion of the mitral valve.

Echocardiographic findings of a LSVC include dilation of the coronary sinus that may protrude into the left atrium and be mistaken for a left atrial mass. A LSVC may be suspected on a chest radiograph if there is a prominent vertical border along the superior mediastinum lateral to the aortic knob. The diagnosis of a LSVC can be confirmed by contrast echocardiography, computerized tomography, or magnetic resonance imaging (Figure 5-Magnetic resonance image angiography demonstrating persistent left superior vena cava.)

Approximately 10% of LSVCs drain into the left atrium. Drainage of a LSVC into the left atrium produces a small right to left shunt. However, most patients with drainage of a LSVC into the left atrium have associated atrial septal defects or heterotaxy syndromes.

Drainage of LSVC into the left brachiocephalic vein, right atrium, and right superior vena cava rarely occur.

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American Board of Pediatrics Content Specification(s):

Know normal and abnormal embryologic development of the heart and great arteries and the factors affecting these

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The analysis of pleural fluid is helpful in the diagnosis of congenital chylothorax. The fluid, in the absence of enteral feeding, typically is amber colored, is rich in lymphocytes (differential count >70%), and may not show an elevated triglyceride concentration. With enteral feeding, especially using milk rich in long-chain triglycerides, the pleural fluid becomes milky in appearance, remains rich in lymphocytes, and shows an elevated triglyceride concentration (>110 mg/dL [1.2 mmol/L]). The evolution of pleural effusion in the infant in this vignette, its manifestation at birth before any postnatal interventions, and the pleural fluid analysis are compatible with the diagnosis of congenital chylothorax.

Pleural effusion resulting from heart failure often is bilateral and accompanied by hydrops. Hydrops fetalis may be associated with congenital structural heart malformations and abnormalities of cardiac rhythm. The structural malformations typically include hypoplastic left heart syndrome and endocardial cushion defect. Among the rhythm abnormalities, tachyarrhythmias, including supraventricular tachycardia and atrial flutter, are more common than bradyarrhythmias such as heart block. The severity and unilateral localization of the pleural effusion as well as the absence of generalized edema make heart failure an unlikely cause of pleural effusion in the infant in this vignette.

Extravasation of fluid into the pleural space can result from injury to the thoracic duct, or obstruction of the subclavian vein or superior vena cava. Surgical procedures, such as correction of coarctation of aorta, ligation of patent ductus arteriosus, and repair of congenital diaphragmatic hernia, may cause inadvertent injury to the thoracic duct. The thrombosis of the subclavian vein or superior vena cava and resultant increase in central venous pressure may cause extravasation of fluid into the pleural space. The thrombosis often is a complication of long-term use of indwelling catheters for administration of parenteral nutrition. The absence of any of these interventions makes iatrogenic extravasation an unlikely cause of pleural effusion in the infant in this vignette.

Pneumonia resulting from perinatally acquired bacterial infection involving organisms such as group B *Streptococcus* may be associated with pleural effusion. Typically, the effusion is bilateral, less voluminous than that described in this vignette, and characterized by neutrophilic preponderance. Rarely, in extreme cases of late-onset bacterial sepsis, the pneumonia may be complicated by a localized collection of pus as in lung abscess or empyema. The nature of the pleural fluid and the absence of features suggestive of sepsis make intrauterine infection an unlikely cause of pleural effusion in the infant in this vignette.

Mediastinal malignancies, such as lymphoma, sarcoma, or neuroblastoma, are rare causes of pleural effusion in neonates. The effusion in such cases may result from obstruction and rupture of the lymphatics induced by the tumor, or from invasion of the lymphatics by the tumor.

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References:

Beghetti M, La Scala G, Belli D, Bugmann P, Kalangos A, LeCoultre C. Etiology and management of pediatric chylothorax. *J Pediatr.* 2000;136:653-658

Devine PC, Malone FD. Noncardiac thoracic anomalies. *Clin Perinatol.* 2000:27:865-899

Dubin PJ, King IN, Gallagher PG. Congenital chylothorax. *Curr Opin Pediatr.*

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Wasmuth-Pietzuch A, Hansmann M, Bartmann P, Heep A. Congenital chylothorax: lymphopenia and high risk of neonatal infections. *Acta Paediatr.* 2004;93:220-224

American Board of Pediatrics Content Specification(s):

Understand the pathophysiology and recognize the clinical, radiographic, and laboratory manifestations of hydrothorax/chylothorax

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January: Question 2

A term male infant with primary persistent pulmonary hypertension of the newborn (PPHN) requires substantial support with fraction of inspired oxygen (Fio_2) of 1.0, high-frequency oscillatory ventilation, dopamine, and inhaled nitric oxide. The mean airway pressure (MAP) on the ventilator is 24 cm H_2O and the partial pressure of arterial oxygen (Pao_2; right radial) is 60 torr. The oxygenation index calculated from these values is 40 (MAP × Fio_2 × 100/Pao_2). After discussion with the infant's parents, venovenous extracorporeal membrane oxygenation (VV ECMO) using a double lumen catheter is initiated.

Of the following, the MOST accurate statement about the expected physiologic effects of VV ECMO is that:

1	Cardiac output is reduced because blood flow to the coronary circulation is decreased.
2	Oxygen consumption is reduced because venous oxygen content is increased.
3	Oxygen extraction increases because systemic arterial oxygen content is increased.
4	Pulmonary blood flow increases because the oxygen content of blood perfusing the pulmonary circulation is increased.
5	Systemic oxygen delivery increases because arterial oxygen saturation is maximized (>98%).

You selected 3, the correct answer is 4.

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Persistent pulmonary hypertension of the newborn (PPHN) affects 1 to 2 per 1,000 live births. Most infants with PPHN are born after 34 weeks' gestation, and have an underlying cardiorespiratory disorder such as meconium aspiration syndrome, sepsis, transient tachypnea, pneumothorax, congenital diaphragmatic hernia, and some congenital heart lesions (eg, transposition of the great vessels, total anomalous pulmonary venous return). Primary PPHN affects about 10% of infants with this illness.

The essential pathophysiology of PPHN involves persistent or recurrent elevation in pulmonary vascular resistance after birth, often associated with hypoxic respiratory failure (Figures 1 and 2).

Figure 1: Normal newborn.

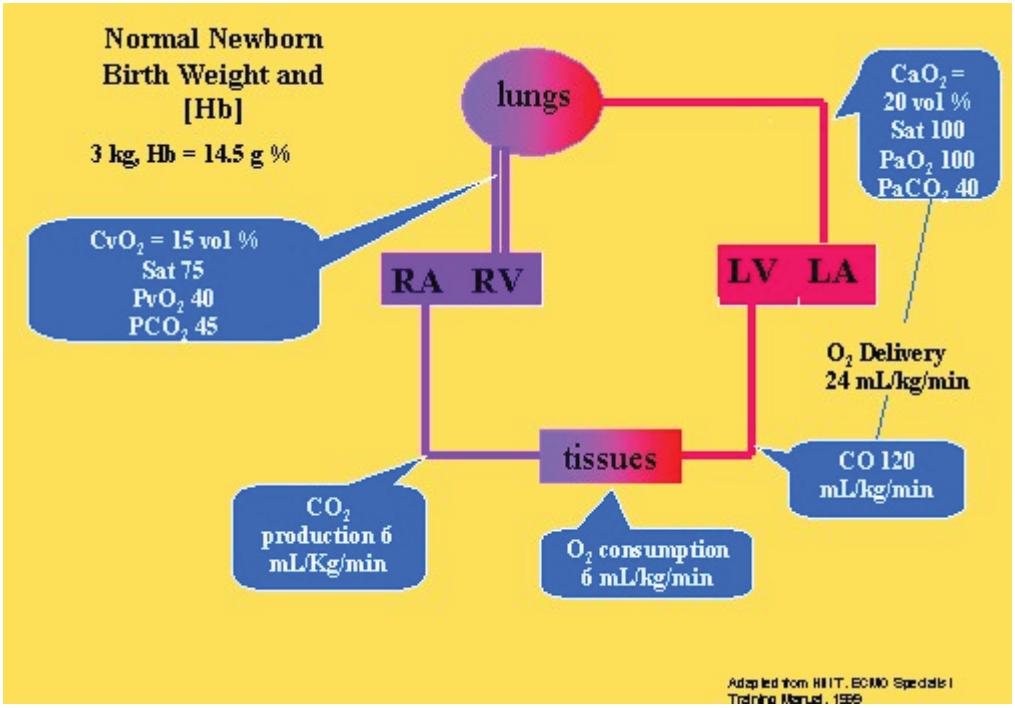
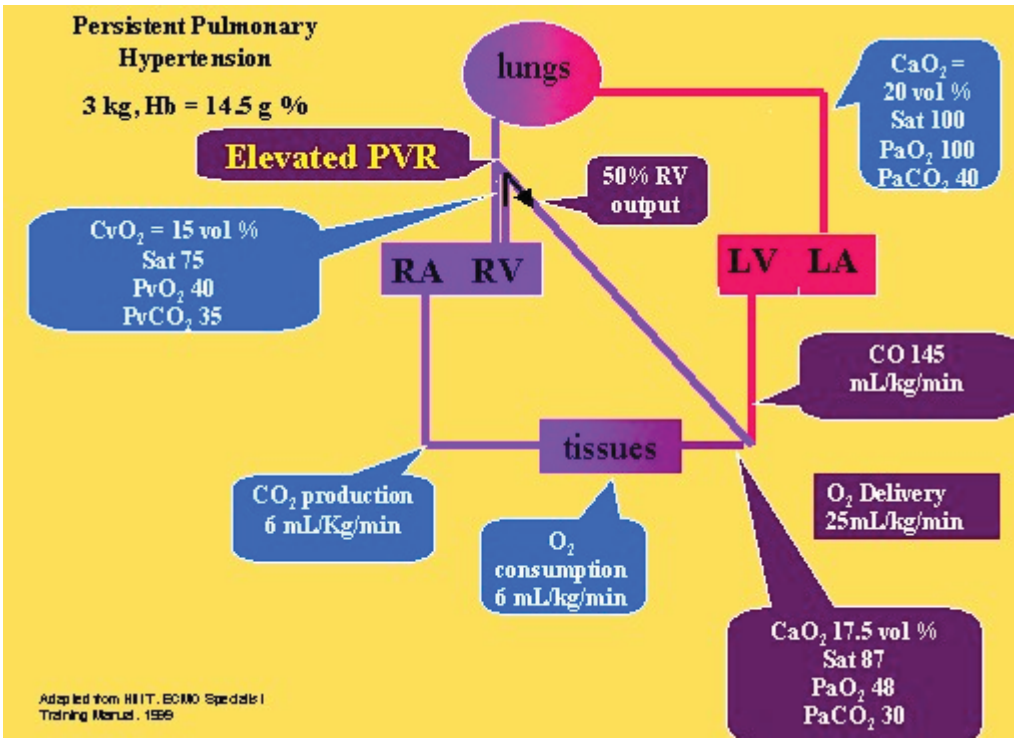


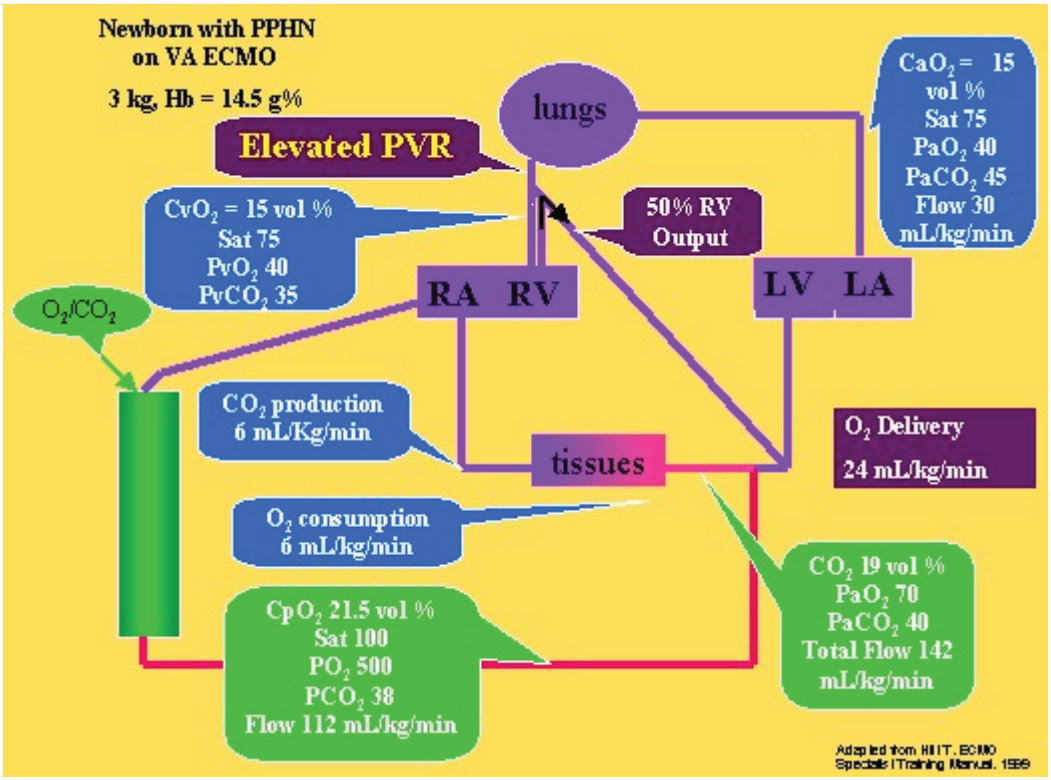
Figure 2: Newborn and persistent pulmonary hypertension (PPHN).



Venous blood is shunted from the right-sided circulation to the left-sided circulation through the ductus arteriosus, foramen ovale, and across poorly functioning diseased lung segments. These right-to-left shunts lead to systemic hypoxemia and cyanosis. Improvements in care have increased survival of such infants. Physiology-targeted management strategies, high-frequency ventilation, surfactant, and inhaled nitric oxide have proven beneficial in clinical series and randomized clinical trials. Extracorporeal membrane oxygenation (ECMO), usually initiated as a rescue intervention when all other treatments have failed to reverse the clinical spiral of PPHN, was also proven beneficial in a multicenter, randomized, controlled clinical trial.

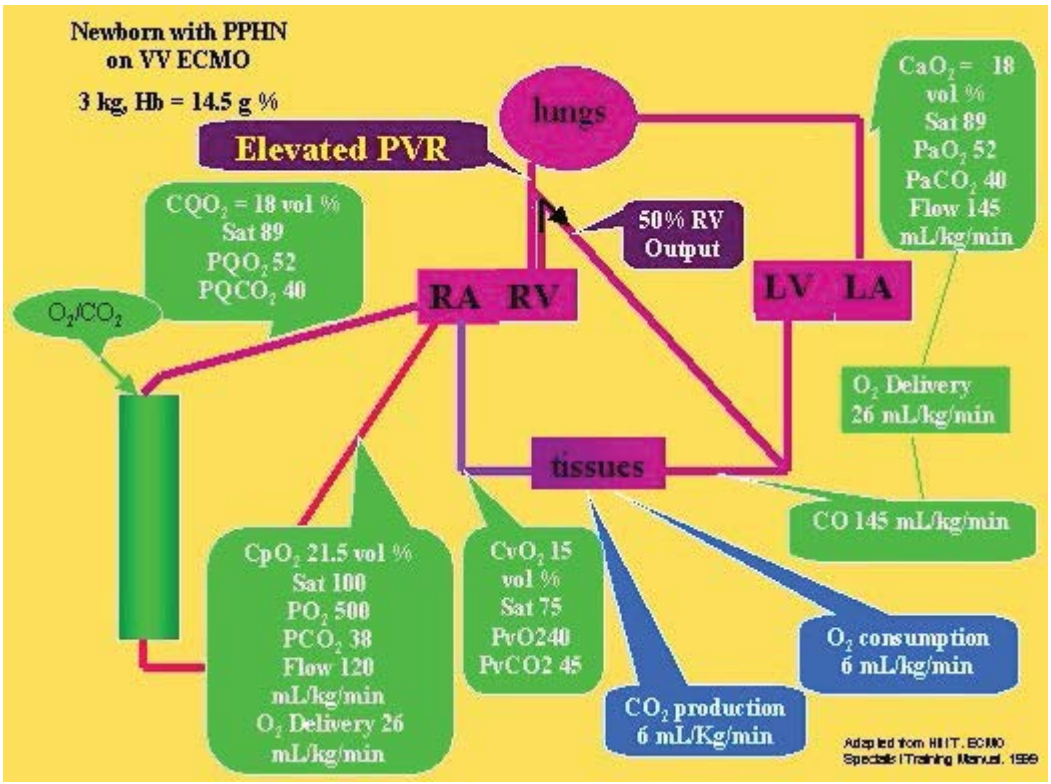
Extracorporeal membrane oxygenation is most often applied to newborns with respiratory failure through catheters placed in both the right internal jugular vein and right carotid artery (venoarterial ECMO or VA ECMO, Figure 3)

Figure 3: Newborn and persistent pulmonary hypertension (PPHN) and venoarterial extracorporeal membrane oxygenation (ECMO).



or through a single double-lumen catheter in the right internal jugular vein (venovenous ECMO or VV ECMO, Figure 4).

Figure 4: Newborn and persistent pulmonary hypertension (PPHN) and venovenous extracorporeal membrane oxygenation (ECMO).



As seen in the infant in the vignette, VV ECMO provides well-oxygenated blood to the pulmonary circulation. Improved pulmonary arterial oxygenation is a potent vasodilator that reduces pulmonary vascular resistance and increases pulmonary blood flow. The mechanism for the oxygen effect is complex, involving vasodilatation and alleviation of hypoxic vasoconstriction. Increased oxygen tension improves blood flow, which induces shear-stress-related vasodilatation, reduces myogenic tone, and increases endogenous production of a number of different endothelial- and nonendothelial-derived vasodilators, especially nitric oxide and prostacyclin.

Venovenous ECMO indirectly improves cardiac function. With increased pulmonary blood flow, pulmonary venous return to the left atrium increases, thereby providing more adequate preload for the left ventricle. Importantly, blood that flows to the coronary ostia and circulation is derived from the left ventricle in both VV ECMO and VA ECMO. During VV ECMO, pulmonary venous blood is highly saturated with oxygen. In contrast, VA ECMO diverts blood from the right atrium and returns it to the aortic arch, essentially bypassing the heart completely at total ECMO support. Blood from the right atrium is venous in origin and the volume of blood flow through the pulmonary circulation to the left atrium and left ventricle is reduced. Thus, during complete VA ECMO support, there is a reduction in cardiac output and oxygen tension that may compromise cardiac function. The higher risk of cardiac stun during VA ECMO compared with VV ECMO is indirect evidence for reduced oxygen delivery to the coronary circulation during VA ECMO. Of note, during VA ECMO the extracorporeal support is most often partial, and oxygen delivery to the coronary circulation is adequate.

Oxygen consumption is calculated by multiplying the cardiac output (CO) by the difference between the oxygen content of the arterial (Cao_2) and venous circulations (Cvo_2):

$$O_2 \text{ Consumption} = CO \times (Cao_2 - Cvo_2)$$

$$O_2 \text{ Content} = \{(Hb) \times 1.34 \text{ mL } O_2/\text{g Hb} \times So_2\} + 0.003 \times Po_2$$

During VV ECMO, O_2 consumption is maintained as long as adequate oxygen delivery is present; it is not reduced. Oxygen delivery (DO) equals the CO

multiplied by the oxygen content (Cao_2).

$$O_2 \text{ Delivery} = CO \times Cao_2$$

As stated before, cardiac output likely increases during VV ECMO. The increase in CO usually compensates for any drop in arterial oxygen content that occurs due to lower arterial oxygen saturation (80% to 95%, see O_2 content equation above), which is sometimes seen after the transition to VV ECMO.

Venovenous ECMO and VA ECMO are both intended to provide improved oxygen delivery by increasing arterial oxygen content and cardiac output. Improved oxygen delivery is expected to reverse the compensatory responses of blood flow redistribution and increased oxygen extraction that occur when tissues sense inadequate oxygen delivery. Oxygen extraction should be reduced, and mixed venous oxygen saturation is anticipated to normalize. If oxygen consumption is increased due to seizures, jitteriness, or septic shock, however, additional oxygen delivery may be necessary. Oxygen delivery can be increased with improved cardiac output and/or higher arterial oxygen content (higher arterial oxygen saturation or hemoglobin). If this increase in oxygen delivery cannot be achieved with maximal ECMO flow, blood flow redistribution to essential organs (brain, heart, adrenal glands) and increased oxygen extraction (O_2 extraction) may be required by the tissues of the body (indicated by a decrease in mixed venous oxygen saturation and venous oxygen content).

$$O_2 \text{ Extraction} = (Cao_2 - Cvo_2)/Cao_2$$

Venovenous ECMO improves oxygen delivery by increasing venous oxygen content, increasing cardiac output and, in many instances, increasing arterial oxygen saturation (Sao_2) and content. However, Sao_2 values of 80% to 95% are expected even during full VV ECMO flow (>120-150 mL/kg per minute) because it is the venous admixture in the right atrium that perfuses the systemic circulation (Figure 4). Thus, increased systemic oxygen delivery is not usually due to maximum Sao_2 values (>98%). If endogenous pulmonary function contributes to oxygen exchange, Sao_2 may be greater than 95%. Of note, the relatively lower Sao_2 values achieved with VV ECMO compared with VA ECMO are often accepted in exchange for a reduction in the pulmonary parenchymal and vascular injury associated with high concentrations of oxygen and positive pressure ventilation.

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Hilt T, Graves D, Zwischenberger JB. ECMO physiology. In: Van Meurs K, ed. *ECMO Specialist Training Manual.* 2nd ed. Ann Arbor, Mich: Extracorporeal Life Support Organization; 1999:19-40

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treatment of PPHN. *J Pediatr.* 1995;126:853-864

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American Board of Pediatrics Content Specification(s):

Understand the various factors affecting oxygen uptake, transport, and delivery, including the blood and circulation

Know the management of persistent pulmonary hypertension

Understand the indications for and techniques of extracorporeal membrane oxygenation (ECMO)

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