Cyanosis (pathophysiology)

Cyanosis can be caused by disorders of the respiratory system, congenital heart defects and various right-to-left arteriovenous shunts.

Cyanosis:

V-Q mismatch:

Cyanosis can be divided into peripheral and central .

Definition [edit | edit source]

Cyanosis **is a dark blue discoloration of the mucous membranes and skin** caused by an increased concentration of reduced hemoglobin or methemoglobin in the blood.

It is best seen:

- on the nail bed
- earlobes,
- mucous membranes,
- wherever the skin is thin.

The presence of > 50 g/l of reduced Hb or > 15 g/l of methemoglobin is required for cyanosis to occur. Clinically, we are able to recognize cyanosis as a rule when SaO 2 drops < 80%.

Forms of cyanosis [edit | edit source]

We fundamentally distinguish two forms of cyanosis: **peripheral** and **central .**

- **Central cyanosis**
  - ↓ SaO2
    - in anemia: indicates a serious decrease in saturation
    - in polyglobulia: it manifests even with a smaller decrease in saturation

- on the trunk and limbs as well as on the mucous membrane of the oral cavity and tongue
- It is most often a manifestation of systemic hypoxemia , methemoglobinemia or severe polycythemia .

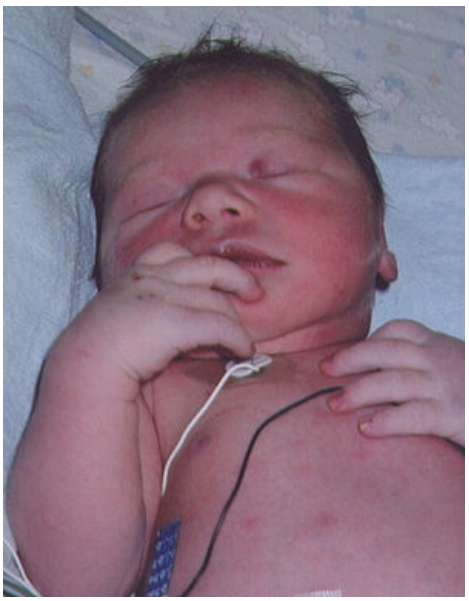
- **Peripheral cyanosis**
  - normal SaO 2 , but ↑a-vO 2 difference (due to ↑ extraction of O 2 and ↑ deoxygenated blood at the venous end of the capillary bed)
  - on the acral parts of the limbs and circumferentially
  - The causes can be benign , eg cold exposure, mild methemoglobinemia
  - In newborns, a few hours after birth, peripheral cyanosis may not even be a pathological symptom - we are talking about neonatal acrocyanosis .
  - Serious causes include septic or cardiogenic shock .

Lack of oxygen is not always accompanied by cyanosis . With a low level of hemoglobin (anemia), a lack of O2, can occur without reaching the concentration of deoxygenated (reduced) hemoglobin necessary for cyanosis. On the contrary, with an increased concentration of hemoglobin in the blood, cyanosis sets in relatively easily, without a lack of O 2 .

Pathophysiology [edit | edit source]

Factors determining the development of cyanosis:

- the absolute amount of hemoglobin in the blood,
- degree of oxygen saturation of hemoglobin,
- qualitative changes in hemoglobin,
- state of circulation.


Cyanotic newborn

Amount of hemoglobin [edit | edit source]

As already mentioned above, the intensity of cyanosis is significantly determined by the absolute amount of reduced Hb. Therefore, with anemia , cyanosis appears only with very severe hypoxemia (or it may not appear at all), on the other hand, with polyglobulia, cyanosis is visible very early and may not be systemic hypoxemia at all .

Binding of oxygen to hemoglobin and changes in hemoglobin structure [edit | edit source]

The degree of hemoglobin saturation is determined by several factors, which include:

- the partial pressure of oxygen in the alveoli,
- state of the alveolocapillary membrane,
- the ability of oxygen to move from the alveoli to the blood,
- the subsequent binding of oxygen to Hb.

If the level of alveolar ventilation decreases, the partial pressure of oxygen in the alveoli decreases, subsequently also in the arterial blood, and desaturation occurs . But even situations with good alveolar ventilation and a physiological value of the partial pressure of oxygen in the alveoli can lead to cyanosis if the transport through the alveolocapillary membrane is disturbed.

Physiologically, oxygen binds reversibly to the Hb iron molecule and thereby changes its structure. Oxygenated Hb is bright red. Factors that affect the binding of oxygen to Hb also affect its color. For example, in carbon monoxide = CO poisoning, there is a competitive binding of CO to the Hb molecule. The binding of CO to Hb is 200 times stronger than that of oxygen. CO bound to the heme group increases the affinity for O 2 of the other three heme groups of hemoglobin, so that the bound O 2 is more difficult to release. Increased affinity for O2 with reduced peripheral O2 output is also present in all conditions that shift the hemoglobin dissociation curve to the left.

The resulting carboxyhemoglobin changes the color of Hb to cherry red. If the iron in Hb is oxidized to the ferric form, known as methemoglobin, it is also unable to transmit oxygen . The blood then has a purple-brown (chocolate) color.

Circulation status

The state of circulation plays a significant role in the intensity of cyanosis:

- cyanosis can be the result of anatomical short-circuits = shunts . A shunt is a situation where deoxygenated blood mixes with arterial blood. If the shunt is significant, pO 2 falls and cyanosis appears. At the pathological level, it is mainly VVV of the heart with a right-to-left shunt.

- cyanosis may be the result of functional shunts due to a mismatch between ventilation and perfusion in the lungs. Perfusion of non-ventilated alveoli gives rise to "pulmonary shunts" and the result is again a drop in pO 2 and cyanosis.

- the third cause of cyanosis due to circulatory disorders is conditions with peripheral hypoperfusion, when slow capillary flow leads to the accumulation of deoxyhemoglobin (cold, shock). Cyanosis is especially noticeable on

the hands, feet and periorally. Cold causes a slow transit of erythrocytes through the capillary, so tissues take up more oxygen. Patients in septic or cardiogenic shock are cyanotic due to prolonged capillary refill time. The cause is vascular collapse in sepsis or heart pump failure.

Differential diagnosis [edit | edit source]

The most common causes of cyanosis:

- local cyanosis;
- generalized cyanosis;
- acrocyanosis of newborns;
- respiratory dysfunction;
- exposure to cold;
- VVV heart.

Causes of cyanosis [edit | edit source]

- Hypoventilation (often in neurological disorders);
- alveolocapillary block;
- pulmonary PL shunts (functional);
- cardiac PL shunts (anatomical);
- abnormal hemoglobins;
- polycythemia ;
- cold;
- sepsis;
- shock;
- hypoglycemia

Methemoglobinémie

Methemoglobinemia is not very common, but must be considered in the differential diagnosis of cyanosis.

- **congenital ,**
- **obtained** .

Congenital methemoglobinemia is caused by either abnormal hemoglobin M or a deficiency of NADH-dependent methemoglobin reductase. Congenital methemoglobinemias are rare.

More common is the acquired variant, which occurs when hemoglobin is exposed to oxidizing substances. Young children exposed to these substances or during gastroenteritis are more sensitive to the immaturity of the enzymatic systems that allow Hb reduction. Symptomatology includes headache, dizziness, dyspnea , confusion, convulsions , and even coma.

Patient access [edit | edit source]

If cyanosis is detected in the patient, we will provide 100% O 2 administration , we will also provide a line and administration of fluids. Monitoring of vital functions is basic.

First of all, we must rule out the following causes: hypoventilation , lung pathology (V/Q imbalance, pulmonary shunts, alveolocapillary block), VVV heart with PL shunt, abnormal hemoglobins, peripheral circulation disorders (including hypothermia), sepsis, hypoglycemia.

With hypoxemia, we expect stimulation of breathing (tachypnea, dyspnea), with the exception of premature infants, who react with hypoventilation and apnea.

Reduced respiratory effort in a child with hypoxemia most likely indicates CNS depression , a neuromuscular disorder, but the possibility of exhausting respiratory effort in a patient with prolonged respiratory distress must also be taken into account.

In the case of respiratory distress with a simultaneous pathological auscultatory finding (whistles, wheezes, rasps, crunches, weakened breathing), we think first of all of the respiratory cause, but also of cases of congestive heart failure (hypercapnia is often found in heart defects with significant pulmonary congestion).

Evaluation of the respiratory system [edit | edit source]

The clinical border of the upper and lower airways is the junction of the clavicles. Retraction with inspiratory stridor is accompanied by extrathoracic airway obstruction.

Mixed stridor means obstruction of intra- and extrathoracic airways (larynx , trachea).

Wheezing with auscultation of squeaks/whistles accompanies obstruction of the lower = intrathoracic airways. The finding of moist phenomena on the lungs may indicate inflammatory involvement of the lower respiratory tract or congestive heart failure.

Weakened or absent breathing is accompanied by large atelectasis and pathology of the pleural space.

Retraction with grunting means impaired lung compliance and loss of functional residual FRC capacity. Grunting is caused by the glottis clenching mechanism. It is an attempt to increase autoPEEP and thereby increase FRC. The result is the prevention of alveolar collapse during expiration. Alar flexion is a mechanism that reduces airway resistance and thereby reduces the work of breathing. Involvement of the sternocleidomastoid muscle, soft parts of the chest and sub-rib is an effort to increase the respiratory volume in a situation where normal minute ventilation no longer covers the needs of the organism.

Evaluation of the cardiovascular system [edit | edit source]

Tachycardia is usually present as a compensatory mechanism, as an increase in cardiac output leads to an increase in tissue oxygen delivery.

We note the manifestations of right-sided heart failure:

- swelling (especially periorbital)
- hepatomegaly
- jugular vein distension

Left-sided heart failure is manifested by pulmonary congestion (dyspnea, tachypnea, auscultation of crackles), cold periphery, harder-to-palpable peripheral pulsations, slowed capillary return, oliguria.

Cyanosis with a predominance of the lower half of the body in a newborn is indicative of coarctation of the aorta, cyanosis with a predominance of the upper half of the body in a newborn is indicative of TGA with an open duct.

Cyanosis affecting, for example, one limb is caused by a disorder of the distal vascular apparatus or the autonomic system. We see it in traumas affecting, among other things, blood vessels or in reflex sympathetic dystrophy.

Evaluation of other systems [edit | edit source]

Within the framework of the CNS, we evaluate:

- level of consciousness
- we rule out head and spinal cord trauma,
- we will assess muscle tone and strength.

From the point of view of the gastrointestinal system, abdominal distension of any origin worsens gas exchange parameters. A boat-shaped abdomen may indicate a diaphragmatic hernia (prenatal ultrasound and postnatal X-ray of the chest and abdomen are most informative in this case). Intermittent cyanosis can be seen, for example, in patients with GER. Apneic pauses with desaturation and cyanosis are typical of newborns and infants with pertussis.

Assessment and diagnosis [edit | edit source]

In a cyanotic child, but without alteration of the general condition with normal pO 2 , it is most often a non-urgent condition. Polycythemia, mild methemoglobinemia, cold exposure, neonatal acrocyanosis , skin problems come into consideration . A KO+dif examination would be sufficient within the laboratory. and methemoglobin.

Despite a normal pO 2, children with altered status need a comprehensive examination very urgently. The cause may be severe methemoglobinemia, carboxyhemoglobinemia, septic or cardiogenic shock. We perform blood counts, blood tests for abnormal hemoglobins, complete biochemical tests including acid-base balance. On the X-ray of the chest, we can find lung pathology or the expansion of the heart shadow. With normal findings, neurological causes, obstruction of the upper airways must be ruled out. A heart murmur during a physical examination leads to the consideration of heart disease, so we supplement the ECG and echocardiography.

As part of the laboratory examination, we determine hemoglobin and hematocrit. In methemoglobinemia, a drop of blood on filter paper turns chocolate brown after a few minutes.

In carbon monoxide poisoning, the pulse oximeter ignores pathological Hb molecules and only detects oxyHb => falsely normal SaO 2 values . In methemoglobinemia, metHb has the same absorption capacity for red and infrared light. This results in a false SaO 2 value of 85%. Both carboxyhemoglobinemia and methemoglobinemia will have normal pO 2 values , as pO 2 reflects physically dissolved oxygen and therefore does not depend on the quality of hemoglobin.

In newborns, the so-called test according to Moore helps in the differential diagnosis of cyanosis: after a 10-minute inhalation of 100% O 2 , in respiratory causes of cyanosis, pO 2 rises > 20 kPa (> 150 torr), in cardiac etiology, pO 2 remains < 14 kPa (< 100 torr). When measuring SaO2 preductally (right hand) and postductally (lower limbs), a difference > 10% indicates a significant P – L shunt across the ductus in the diagnosis of persistent pulmonary hypertension in newborns with PPHN.

Cyanosis caused by alveolar hypoventilation responds well to oxygen supplementation and the cyanosis subsides. In conditions with impaired alveolocapillary transport or in conditions with a significant V/Q disparity, the response to O 2 administration is inadequate.

A-a gradient:

Links [edit | edit source]

Related Articles [edit | edit source]

- Peripheral cyanosis
- Central cyanosis

References [edit | edit source]