HYPERCYANOTIC SPELLS

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Introduction

The 'Tet spell' (also called 'hypoxic spell', 'cyanotic spell', 'hypercyanotic spell', 'paroxysmal dyspnea') is an episodic central cyanosis due to total occlusion of right ventricle outflow in a patient with a congenital heart disease, such as Tetralogy of Fallot (TOF). Hypercyanotic spell characterized by paroxysm of hyperpnea (rapid and deep respirations), irritability and prolonged cry, increased cyanosis and decreased intensity of heart murmur. If not treated in time it may lead to limppness, seizures, neurological deficit and death. A spell is most likely to be seen a child less than 2 year old, upon waking up in the morning and following a crying episode (1). Weng YM et al reported cyanotic spell in a 29-year-old man of TOF also (2).

Pathophysiology

In TOF, The level of cyanosis and onset of cyanotic spell is determined the SVR & level of PS component. In case of mild PS, the RV pressures are usually less than the left ventricle and hence the shunt is usually left to right. If Severe PS then the RV after load becomes high and hence the RV pressures become high. If the SVR is low (TOF with cyanotic spells) then the shunt flow becomes right to left. This results in progressive cyanosis. The resulting fall in arterial PO2 in addition to an increase in PCO2 and fall in pH stimulate the respiratory centre à Increased rate and depth of breathing i.e. hyperpnoea à increase in the systemic venous return to the RV à In the presence of fixed resistance at the right ventricular outflow tract or decreased SVR, the increased systemic venous return to the RV must go out the aorta à Further decrease in the arterial oxygen saturation à Right to left shunt of deoxygenated blood, thus leading to a vicious cycle of hypoxic spells (fig 1).

Figure 1. Mechanism of Hypoxic Spell

Five mechanisms are involved in the pathogenesis of fallot spells 1) An acceleration in heart rate 2) An increase in cardiac output and venous return 3) An increase in right to left shunt 4) Vulnerable respiratory control centers and 5) Infundibular contraction. Manual compression of abdominal aorta can abort spell by decreasing cardiac output and decreasing cardiac output (3-4).
Certain theories have also been postulated as possible explanation for the cause of cyanotic spells. These theories have basically compared the onset of cyanotic spells to exercise in normal individuals where there is fall in systemic arterial oxygen saturation during exercise and which reverses once exercise stops. In cyanotic spells this process of low systemic oxygen saturation continues and ultimately leads to progressive metabolic acidosis. These theories are: Woods et al(5) reported that hypoxemic spells are caused by spasm of the infundibulum of the RV, a progressively increasing right to left shunting and metabolic acidosis. Surge in Catecholamine release leads to increased myocardial contractility and infundibular stenosis. Guntheroth et al(6) reported that episodes of paroxysmal hyperpnea are the cause rather than the effect of cyanotic spells. Hyperpnea increases the systemic venous return leading to right to left shunt as well as oxygen consumption through increase work of breathing. Kothari SS(7) argued against the views mentioned previously and suggested the role of stimulation of mechanoreceptors in the RV to be the cause of spells. Increased contractility (due to catecholamine) and decreased right ventricular size (due to various factors) can trigger a reflex resulting in hyperventilation, some peripheral vasodilation without bradycardia, and this may initiate a spell.

**Clinical Manifestation**

Typically cyanotic spells occur early in the morning. The possible triggers are anxiety, fever, anemia, sepsis or even spontaneously without any cause. The spell are typically initiated by the stress of feeding, crying or bowel movement, particularly after an infant awakens from a long deep sleep. A typical infant with cyanotic spell would appear fussy, inconsolable and thereafter progresses to increasing cyanosis, hyperpnea that is typical of a spell. The older child experiencing a hypoxic spell will often squat to recover. Squatting compresses the superior vena cava and increases systemic vascular resistance, directing blood through the pulmonary stenosis and into the lungs.

Cardiac causes other than TOF that may present with cyanotic spells are tricuspid atresia with pulmonary stenosis (PS), Transposition of great vessels with PS, Single ventricle physiology with PS or pulmonary atresia.

**Management**

*Squatting* / *Knee-to-chest* : Placing the child in the knee-chest position either lying supine or over the parent’s shoulder. This calms the infant, reduces systemic venous return and increases systemic vascular resistance. Various postures (1) assumed for relief of dyspnoea (fig2).

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**Figure 2.** Various postures assumed for relief of dyspnea in TOF 1) squatting, 2) sitting with legs drawn underneath (squatting equivalent), 3) legs crossed while standing, 4) infant held with legs flexed on its abdomen, and 5) lying down.
There is very limited benefit to administer oxygen, since the problem is reduced pulmonary blood flow, not the ability to deliver oxygen to the lungs. Administer morphine sulfate 0.1 mg/kg IV or IM. It depresses respiratory center and lead to decreases in systemic venous return.

**Correct acidosis**: Obtain pH, give Inj. Soda bicarbonate (1-2 meq/kg IV). It reduces the respiratory stimulation by metabolic acidosis, and may diminish the increase in pulmonary vascular resistance caused by hypoxia and acidosis. Propranolol, 0.1 mg/kg slow IV push. May be repeated in 15 minutes. By decreasing cardiac contractility, propranolol may decrease infundibular obstruction of right ventricular outflow. Given orally at 2-4 mg/kg/day PO to prevent spells. When used chronically, have the beneficial effect of stabilizing peripheral vascular reactivity. or Inj Esmolol (0.5mg/kg over 1 min then 50mcg/kg/min over 4 min. or Inj Metoprolol- 0.1mg/kg over 5 min, repeat every 5 min to max 3 doses , then start infusion 1-5 mcg/kg/min. Phenytoin 5-20 mcg/kg IV every 10-15 minutes. Increases the SVR, forcing more blood flow to the lungs. Continuous phenylephrine infusion to maintain adequate pulmonary blood flow to keep oxygen saturations in the 90.

A phenylephrine drip may be run at 0.1-0.5 mcg/kg/min, titrated to desired effect. It is a potent vasoconstrictor that will result in reduced renal and mesenteric perfusion as well. Ketamine- 0.25 - 1.0 mg/kg, IV or IM? has dual benefit causes sedation and increase SVR. Methoxamine - 0.10mg/kg IV over 5-10 min. --> Leads to increase SVR.

**Intravenous fluids** - preferably initially as bolus of 10-20cc/kg à 60cc/kg. Crystalloid or colloid fluid bolus: This maximises preload and should be given prior to the following drugs which may induce hypotension. Correct anemia and consider operation.

References
8. Park MK. Pediatric cardiology for Practitioners. 4th edn. Mosby, St. Louis 2004: 123