

Indian Academy of Pediatrics (IAP)



# STANDARD TREATMENT GUIDELINES 2022



## Cyanotic Spell

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**Under the Auspices of the IAP Action Plan 2022**

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# Cyanotic Spell

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## Cyanotic Spells

### Introduction

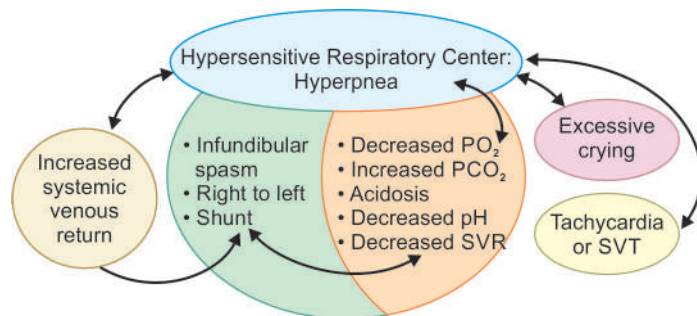
- ☑ Cyanotic spells (CSs) are paroxysmal episodes of deepening of cyanosis, hyperpnea, excessive crying, tachycardia, and exhaustion culminating into sleep, or unconsciousness in infants and toddlers who are born with cyanotic congenital heart disease (CCHD) comprised of decreased pulmonary blood flow and large ventricular septal defect [tetralogy of Fallot (TOF) or CCHD with TOF physiology].
- ☑ In a few cases, these spells may culminate into seizures and death.
- ☑ These episodes are attributed to intermittent infundibular spasms, increased right ventricular outflow obstruction (RVOTO), increased right-to-left shunting, metabolic acidosis, stimulation of central respiratory centers, and hyperventilation or hyperpnea.

- ☑ Cyanotic spell happens in approximately 40% of infants and young children with TOF physiology. The onset of CS is usually in the early morning after several hours of sleep.
- ☑ Routine activities such as feeding, crying, or bowel movements or disease states such as infection and anemia may also precipitate it.

### Precipitation of Cyanotic Spell

Tetralogy of Fallot and Similar Congenital Heart Diseases  
with Reduced Pulmonary Blood Flow and Large  
Interventricular Connection (Fig. 1)

- ☑ The usual substrate for CS is the classical tetralogy of Fallot which is a combined term for ventricular septal defect (VSD), malaligned outlet septum, overriding aorta, severe pulmonary stenosis (PS), and right ventricular hypertrophy.
- ☑ Hyperpnea in TOF causes increased pulmonary vascular resistance and adds up to impedance offered by the already obstructed right ventricular outflow tract during CS. It also increases the oxygen requirement but due to fixed RVOTO, the compensatory increase in systemic venous return cannot enter in lungs, and eventually gets redirected to the aorta due to the presence of large VSD, an exit with low impedance, leading to an exaggeration of systemic hypoxia, acidosis, and vasodilatation.



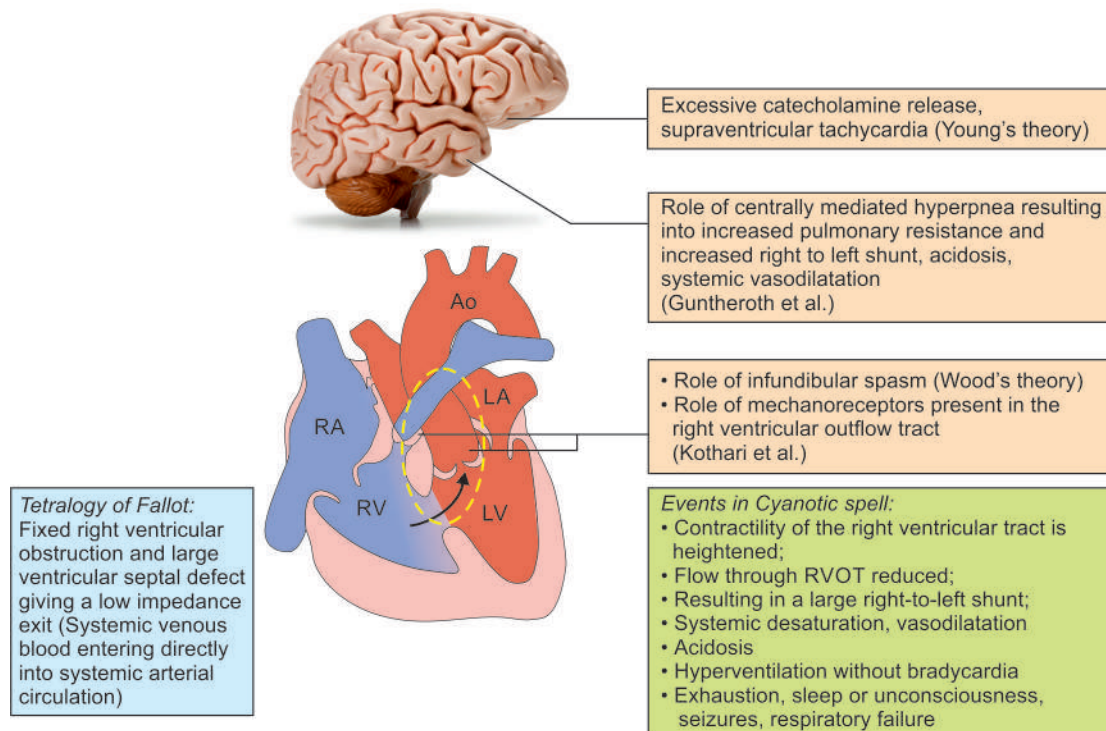
**Fig. 1:** Pathophysiology of cyanotic spell: Vicious cycle of catastrophic changes.

- ☑ VSD and pulmonary atresia (PA)
- ☑ Double outlet right ventricle (DORV), VSD, PS, or PA
- ☑ Single ventricle physiology (double inlet right or left ventricle or tricuspid atresia), VSD, and PS/PA
- ☑ Complete transposition of great vessels (D-TGA), VSD, and PS/PA
- ☑ Corrected transposition of great vessels (cTGA), VSD, and PS/PA

Cyanotic Congenital Heart  
Disease with Tetralogy of  
Fallot Physiology

Paroxysmal nature of CS remains unexplained and therefore multiple theories have been proposed as the perpetrator CS (**Fig. 2**):

- ✓ **Wood's Theory:** Increased infundibular spasm leads to the increased right-to-left shunting across the VSD and leads to hypoxia and subsequent catastrophic events.
- ✓ **Guntheroth and Morgan Theory:** Hyperpnea probably precipitated by hypersensitive respiratory centers and tachycardia leads to increased oxygen requirement, increased cardiac output, increased systemic venous return, increased right-to-left shunt, and culminates into the full-blown CS.
- ✓ **Theory by Young et al.:** Excessive catecholamine release, tachycardia, or supraventricular tachycardia precipitate CS.
- ✓ **Hamilton et al.:** Decreased systemic resistance leads to the increased right-to-left shunt and eventual CS.
- ✓ **Hein's Theory:** Metabolic acidosis as the cause of decreased systemic vascular resistance (SVR) and precipitation of hyperpnea
- ✓ **Kothari SS:** Role of mechanoreceptor found in RV, as the starter of increased RVOTO and increased right-to-left shunting.



**Fig. 2:** Paroxysmal episode of cyanotic spell: Theories.

**TABLE 1:** Evaluation and work-up.

Evaluation	<ul style="list-style-type: none"> <li>☑ SpO<sub>2</sub>, sensorium, respiration, heart rate, temperature dehydration, etc.</li> <li>☑ Review the previous medical records if they are available</li> </ul>
Primary tests	<ul style="list-style-type: none"> <li>☑ SpO<sub>2</sub> by a pulse oximeter</li> <li>☑ X-ray of the chest</li> <li>☑ Electrocardiogram (ECG) (rule out supraventricular tachycardia)</li> <li>☑ Echo</li> <li>☑ Arterial blood gas analysis (after the baby is stabilized)</li> <li>☑ Blood sugar</li> </ul>
Additional tests	<ul style="list-style-type: none"> <li>☑ Rule out anemia (hemoglobin and complete blood count)</li> <li>☑ Rule out infection if fever is present</li> <li>☑ C-reactive protein and blood culture</li> <li>☑ Renal function test, electrolytes</li> </ul>

Diagnosis of the CS is clinical. However, a detailed workup is done to diagnose the exact underlying pathoanatomy, metabolic abnormalities, and to recognize the precipitating factors such as anemia and infection.

- ☑ Paroxysmal crying (period of inconsolable crying)
- ☑ *Hyperpnea*: Characterized by deep breathing with normal lungs
- ☑ Deepening of cyanosis
- ☑ Decreased heart murmur
- ☑ Exhaustion culminates in sleep, unconsciousness, convulsions, and rarely in death (**Figs. 3A to C**).



**Figs. 3A to C:** Cyanotic spell: (A) Irritable inconsolable baby; (B) Exhausted baby after an episode inconsolable of crying; (C) Nebulized oxygen to an irritable child.

Goals of management are as follows:

- ☑ To reduce anxiety (calming down) pain, and therefore to reduce the heart rate and systemic vasodilatation
- ☑ To follow an algorithm-based management plan
- ☑ Volume replacement and correction of acidosis and anemia
- ☑ To control the heart rate and decrease the sympathetic drive with beta-blockers and alpha-1 adrenergic receptor agonists
- ☑ Treatment of arrhythmias, fever, and anemia
- ☑ Long-term management plan and parental counseling in patients who show good recovery
- ☑ Catheterization or surgical intervention for nonresponders

**TABLE 2:** Summary of management (Flowchart 1).

Nonpharmacological therapy	<ul style="list-style-type: none"> <li>☑ Triaging/hospitalization</li> <li>☑ Knee-chest position on the bed or positioning a baby on the lap of the mother or squatting, to reduce venous return and increase the SVR (<b>Figs. 4A to C</b>)</li> <li>☑ Oxygen inhalation (nasal cannula or mask/nebulized form is better and causes less irritation)</li> </ul>
Drugs/fluid	<p><i>The first line of drugs:</i> Preferred sedative + <math>\beta</math>-blockers</p> <ul style="list-style-type: none"> <li>☑ Morphine 0.1 mg/kg (subcutaneous or intramuscular)</li> <li>☑ <i>Injection metoprolol:</i> 0.1 mg/kg slow IV, 3–5 <math>\mu</math>g/kg/min—continuous infusion</li> <li>☑ <i>Ketamine:</i> 0.25–1.0 mg/kg IV or IM</li> </ul> <p><i>The second line of drugs (alternative drugs):</i></p> <ul style="list-style-type: none"> <li>☑ <i>Beta-blockers:</i> <ul style="list-style-type: none"> <li>– <i>Propranolol:</i> 0.1–0.2 mg/kg intravenously over 5 minutes OR</li> <li>– <i>Esmolol</i> (0.5 mg/kg over 1 minute then 50 <math>\mu</math>g/kg/min over 4 minutes)</li> </ul> </li> <li>☑ <i>Alpha-1 adrenergic receptor agonists:</i> <ul style="list-style-type: none"> <li>– <i>Phenylephrine hydrochloride:</i> 0.01 mg/kg IV (slowly) or 0.1 mg/kg SC or IM</li> <li>– <i>Methoxamine:</i> 0.10 mg/kg IV over 5–10 minutes</li> </ul> </li> </ul>
Volume replacement	<ul style="list-style-type: none"> <li>☑ <i>Normal saline infusion (bolus 10–15 mL/kg then maintenance fluid):</i> Fluid restriction is not needed in children with reduced pulmonary blood flow.</li> <li>☑ <i>Blood transfusion (packed red blood cells):</i> 5–10 mL/kg IV over 5 hours (Anemia Hb &lt;12 g% may trigger a cyanotic spell)</li> </ul>
Acidosis	<ul style="list-style-type: none"> <li>☑ <i>IV NaHCO<sub>3</sub>:</i> (1–2 mEq/kg) intravenously slowly can be given to correct metabolic acidosis</li> </ul>
Control of fever	<ul style="list-style-type: none"> <li>☑ Paracetamol (10–15 mg/kg/dose per rectum), IV (to be infused over 15 minutes with appropriate volume)</li> <li>☑ Tepid water sponging must only be attempted in a sedated baby.</li> </ul>

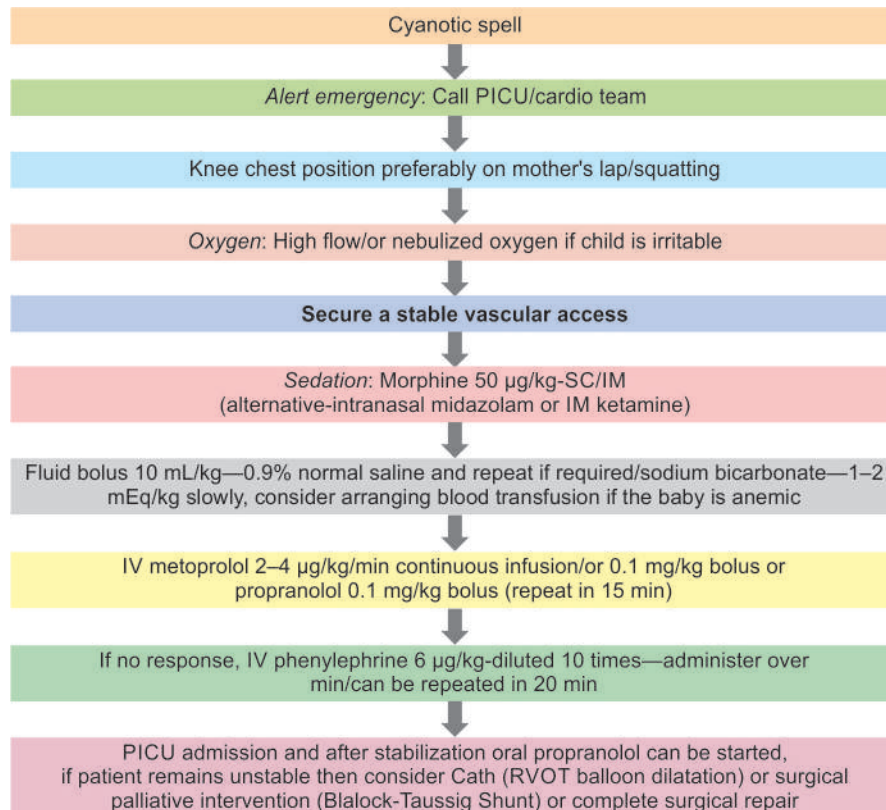
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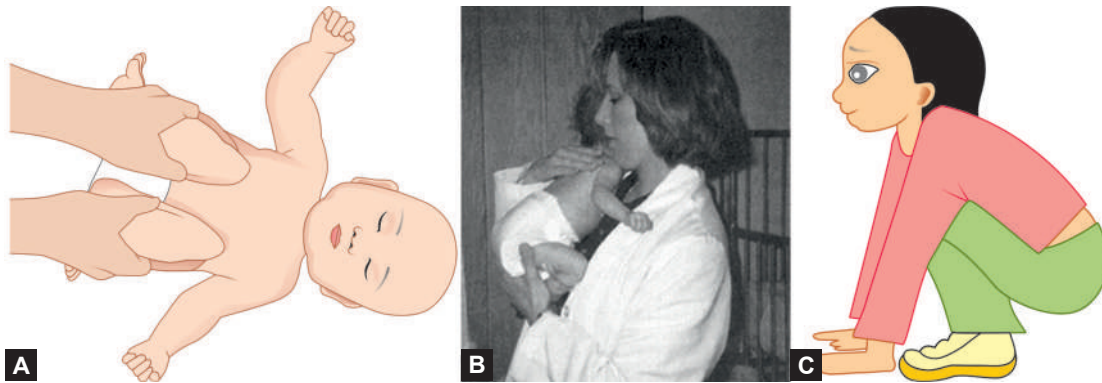
Control of supraventricular arrhythmia	<p><i>Antiarrhythmic drugs:</i> Supraventricular tachycardia may precipitate or sustain the cyanotic spell; therefore, it must be ruled out.</p> <ul style="list-style-type: none"> <li>☑ <i>Adenosine:</i> Initial dose: 0.05–0.1 mg/kg IV bolus over 1–2 seconds</li> <li>☑ <i>Repeat dose:</i> Repeat at incrementally higher doses, increasing by 0.05–0.1 mg/kg, if SVT fails to revert (Refer to Pediatric Cardiologist)</li> <li>☑ Beta-blockers or digoxin (if no WPW syndrome), can be given on a long-term basis</li> </ul>
No responders or presentation with aspiration/respiratory failure	<ul style="list-style-type: none"> <li>☑ Invasive ventilation</li> <li>☑ <i>Cath intervention:</i> Right ventricular outflow stenting</li> <li>☑ <i>Palliative surgical intervention:</i> Blalock–Taussig shunt</li> <li>☑ Corrective surgery (if anatomy is suitable)</li> </ul>
Prevention of cyanotic spell	<ul style="list-style-type: none"> <li>☑ Tab propranolol (1–4 mg/kg/day) oral daily dose divided into two to four doses</li> <li>☑ Iron supplement (maintenance dose if Hb is &gt;13g%)</li> </ul>
Parental counseling and education	Parents must be educated about the disease and instructed about medicines, and their side effects. They must take precautions during episodes of fever or infections to avoid dehydration and seek medical help as early as possible.

(Hb: hemoglobin; IM: intramuscular; IV: intravenous; SVR: systemic vascular resistance; SVT: supraventricular tachycardia; WPW: Wolff–Parkinson–White)

**Flowchart 1:** Algorithm of management of cyanotic spell.







**Figs. 4A to C:** (A) Knee-chest position; (B) Knee-chest position in mother's lap; (C) Squatting.

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- ☑ Shinebourne EA, Anderson RH. Fallot's tetralogy. In: Anderson RH, Baker EJ, Macartney FJ, Rigby ML, Shinebourne EA, Tynan M (Eds). *Paediatric Cardiology*, 2nd edition. London/Toronto: Churchill Livingstone; 2002. pp. 1213-502.