

## HYPERCYANOTIC SPELL

### Introduction

Sudden severe episodes of intense cyanosis caused by reduction of pulmonary flow in patients with underlying Tetralogy of Fallot or other cyanotic heart lesions. This is due to spasm of the right ventricular outflow tract or reduction in systemic vascular resistance (e.g. hypovolaemia) with resulting increased in right to left shunt across the VSD.

### Clinical Presentation

- peak incidence age: 3 to 6 months
- often in the morning, can be precipitated by crying, feeding or defaecation
- severe cyanosis, hyperpnoea, metabolic acidosis
- in severe cases, may lead to syncope, seizure, stroke or death
- there is a reduced intensity of systolic murmur during spell

### Management

- treat this as a *medical emergency*
- knee-chest/squatting position:
  - place the baby on the mother's shoulder with the knees tucked up underneath.
  - this provides a calming effect, reduces systemic venous return and increases systemic vascular resistance
- administer 100% oxygen
- give IV/IM/SC morphine 0.1 – 0.2 mg/kg to reduce distress and hyperpnoea

If above measures fail:

- give IV Propranolol 0.05 – 0.1 mg/kg slow bolus over 10 mins
- alternatively, IV Esmolol 0.5 mg/kg slow bolus over 1 min, followed by 0.05 mg/kg/min for 4 mins.
  - can be given as continuous IV infusion at 0.01 – 0.02 mg/kg/min.
  - Esmolol is an ultra short acting beta blocker
- volume expander (crystalloid or colloid) 20 ml/kg rapid IV push to increase preload
- give IV sodium bicarbonate 1 mEq/kg to correct metabolic acidosis
- heavy sedation, intubation and mechanical ventilation

In resistant cases, consider

- IV Phenylephrine / Noradrenaline infusion to increase systemic vascular resistance and reduce right to left shunt
- emergency Blalock Taussig shunt

*Other notes:*

- a single episode of hypercyanotic spell is an indication for early surgical referral (either total repair or Blalock Taussig shunt).
- oral propranolol 0.2 – 1 mg/kg/dose 8 to 12 hourly should be started soon after stabilization while waiting for surgical intervention.