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# Management of Tet Spell –an updated Review

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## Introduction

Tetralogy of Fallot is the most common cyanotic congenital heart disease. Tet spell, also called a hyper cyanotic spell, hypoxic spell, or cyanotic spell, is a hallmark clinical manifestation of Fallot physiology characterized by a paroxysmal episode of hypoxia due to reduction in pulmonary blood flow [1]. Although typical of Tetralogy of Fallot, other congenital heart diseases with Fallot physiology like double outlet right ventricle with pulmonary stenosis, pulmonary atresia with ventricular septal defect, tricuspid atresia with pulmonary stenosis, and transposition of great arteries with ventricular septal defect and pulmonary stenosis can develop cyanotic spells. With time, the morbidity and mortality associated with Tetralogy of Fallot have improved markedly owing to early detection, corrective surgery and medical care.

## Clinical Feature

The incidence of Tet spell peaks between the second month and second year of life and reduces in frequency after that [2]. Tet spells are exceedingly rare in adults [3]. A typical Tet spell is triggered by a cry, with progressive tachypnea, deep breathing, progressing to worsening of cyanosis, and if not corrected, may end as syncope, convulsions, cerebrovascular accident or rarely death [1-4]. Other triggers are the stress of feeding, waking up from a long deep sleep, fever, dehydration, defecation, anaesthetic agents, cardiac catheterization or rarely supraventricular tachycardia [1-6]. The oxygen saturation is lower than the child's usual levels, and the murmur of right ventricular outflow obstruction softens or disappears.

## Mechanism of Tet Spell

Various triggers cause sympathetic stimulation and catecholamine release that causes contraction of the right ventricular outflow tract (infundibulum). There will be an increase in shunting of deoxygenated blood into the systemic circulation. This is the most accepted mechanism thought to be responsible for further unfavourable hemodynamic changes downstream [4]. Another popular explanation is the presence of a vulnerable respiratory centre, which after a prolonged deep sleep, abnormally responds to sudden increased demand of cardiac output triggered by crying, feeding, or straining [1-5]. Irrespective of the underlying mechanism, the trigger sets in a series of physiological changes leading to a vicious cycle. Increased catecholamine release leads to tachycardia and increased cardiac output, augmenting the deoxygenated systemic venous return to the right ventricle. In the presence of a severe obstruction to pulmonary outflow secondary to infundibular contraction, more deoxygenated blood is shunted across the ventricular defect causing reduction of systemic oxygen content. Decreased systemic arterial oxygen content causes acidosis and hypercarbia. Sensitive respiratory centres respond to these changes by increasing the rate and depth of respiration. This change in respiratory pattern, in turn, causes an increase in the venous return to the right ventricle perpetuating the vicious cycle of cardio-respiratory deterioration.

## Objectives of Treatment

The principle objectives during the management of Tet spell are to decrease catecholamine production, increase blood oxygenation, increase systemic vascular resistance, reduce right ventricular outflow obstruction and increase pulmonary blood flow [7].

## Immediate Measures

### Take Care of the Trigger and the Posture

The management begins with alleviation of anxiety and pain, which reduces catecholamine release and hence reduces heart rate, systemic vasodilation and systemic oxygen consumption. Most of the time, the spell is initiated by the baby's cry; therefore, it is essential that the baby is picked up and comforted, which reduces the sympathetic activity and reduces the PVR. The child should be held in one of many positions known to compress the femoral arteries, increase systemic vascular resistance, and reduce venous return. The most established of these postures being flexion at the knees and hip so that the knees tend to touch the baby's chest. Most of the hyper cyanotic spells are mild and settle with these measures. Squatting is the characteristic posture an older child learns to adapt to terminate the spell [8]. Manual compression of the abdominal aorta in a child has also shown to terminate the episode by a similar mechanism [9]. Various other postures known to be beneficial ('squatting' equivalents) are the knee-chest position, lying down, and sitting with legs drawn underneath. Mild cases of Tet spell terminate with calming and postural manoeuvres [10].

## Supplemental Oxygen

If no improvement is observed, the child needs to be immediately shifted to a hospital facility. Supplemental oxygen should be administered using a face mask or nasal cannula. This increases the oxygen content of the blood and reduces the pulmonary vascular resistance. Meanwhile, intravenous access has to be obtained for further interventions.

## Fluid Administration

Fluid bolus should be administered to increase intravascular volume. Dextrose normal saline can be given as 10ml/kg bolus.



Along with the increase in cardiac output and mixed venous saturation, fluid repletion reduces the risk of hypotension caused by the administration of other drugs. Excessive fluids can lead to cerebral edema, pulmonary edema and hypoxia due to the dilutional effects in the setting of compensatory polycythemia.

### Sedation

Morphine alleviates pain and anxiety, reducing the heart rate and respiratory rate. It decreases the venous return, reduces the pulmonary vascular resistance, and reduces catecholamine release, improving infundibular spasm. Morphine should be given at the dose of 0.1mg/kg to 0.2 mg/kg by intravenous route. The intramuscular or subcutaneous route can be used if intravenous access is not available. Some reports suggest use of midazolam, dexmedetomidine and fentanyl [11-13].

### Beta-Blocker

Administration of parenteral beta-blockers helps in reducing heart rate, improves cardiac filling, preload and may increase systemic vascular resistance. Commonly used drugs are intravenous propranolol at the dose of 0.015 mg/kg to 0.02 mg/kg or short-acting esmolol at the dose of 0.5mg/kg given over one minute [14]. Intravenous metoprolol can be given at the dose of 0.1mg/kg slowly over 5 minutes, and it can be repeated after 5 minutes [maximum 3 doses] followed by infusion at 1-2mcg/kg/min.

### Sodium Bicarbonate

Persistent hypoxia and shock lead to acidosis, contributing to the vicious hemodynamic cycle of hyper cyanotic spell. If the child develops worsening of acidosis, intravenous sodium bicarbonate can be administered at the dose of 1mEq/kg.

### Systemic Vasopressors

Systemic vasoconstriction reduces the right to left shunt and improves pulmonary blood flow [15]. Phenylephrine boluses at dose of 0.005 mg/kg to 0.01mg/kg or norepinephrine infusion at the rate of 0.05 mg/kg/min to 1 mg/kg/min can be given for this purpose. Other options include methoxamine [0.1-0.2 mg/kg/dose IV].

### Advanced Measures

Sedation and mechanical ventilation may be required in extreme cases when the spell is refractory to the above measures. Sedation and mechanical ventilation reduces the work of breathing and reduces systemic oxygen consumption. If the child develops convulsions, diazepam 0.2mg/kg or midazolam 0.1-0.2mg/kg can be given intravenously.

### Immediate Endovascular or Surgical Measures

If the hypoxic spell continuous despite the above measures, the child has to be taken up of palliative modified Blalock-Taussig shunt or definite repair with relief of right ventricular outflow obstruction if feasible. The probable role of manual systemic-to-pulmonary artery auto transfusion for a refractory cyanotic spell has been demonstrated in a case report [16]. Percutaneous stenting of the right ventricular outflow tract has been described as an alternative strategy for emergently improving pulmonary blood flow in Tet spell so that surgery can be undertaken later once the child attains a favourable state[17-18].

### Long Term Measures

Once the cyanotic spell has resolved, a comprehensive neurological assessment must be performed to look for any focal neurological deficits. Propranolol given orally at the dose of 0.25 to 1 mg/kg thrice a day reduce the recurrence of spells and can be used if definitive surgery is delayed [19 to 21]. Parents need to be educated regarding the recurrent nature of the spell and measures to avoid precipitating factors. A small observation has demonstrated the role of cerebral oximetry for the early detection of cyanotic spells during the perioperative period [22]. Children with Tet spells are observed to have a higher prevalence of iron deficiency compared to those without. This may indicate a potential role of iron repletion in decreasing the spells [23]. Mental retardation, cerebral venous sinus thromboses, and nasal speech [velopharyngeal insufficiency] are well known long-term adverse effects of recurrent and prolonged spells. Timely corrective surgery completely abolishes spell and prevents the long-term complications of the cyanotic spells.

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