Treatment of Cyanotic Spells.

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Cyanotic spells occur, most commonly, in infants and children with Tetralogy of Fallot. The genesis of spells is a sudden spasm of the Infundibulum of the right ventircle. Many other congenital cardiac malformations, in which infundibular stenosis is a component, are also prone to cyanotic spells. Even children with pulmonary atresia, occasionally, have cyanotic spells. These are very dramatic and fightening to the parents and physicians.

Recognition :

1. Deep cyanosis of the lips and nails and clubbing of finger nails and toe nails may be present. 2. Deep and rapid breathing. 3. Absent or reduced cardiac murmur; check the records and find the previous Cardiac diagnosis if any. Do not wait for the E.C.G. or X-Ray chest film before starting the treatment.

Management :

1. Keep the patient up-right with legs drawn in a knee-chest position. 2. Keep in 100% oxygen in a hood or mask. 3. Give morphine sulphate 0.1 mg/kg subcutaneously stat. In those patients who have previous history of spells wait for 10 min. prior to giving the morphine, at times spells will settle spontaneously. 4. If a spell is resolving then:— (a) Respiration becomes less distressed. (b) Cardiac murmur becomes audible or is louder than at the beginning of the observation. 5. If improvement continues, leave the child in oxygen undisturbed. 6. If spell continues for more than 30 minutes without improvement. (a) Draw blood for arterial blood gases i.e. pH, $PaCo_2$, and $Pa0_2$ (Note in a prolonged spell PH may be reduced with marked reduction of $PaC0_2$. In early spell $paco_2$ may be elevated suggesting the need for mechanical ventilation). (b) Set up an intravenous D5W + 1/5 saline drip. (c) Give Sodium Bicarbonate ($NaHCo_3$), one mEq/kg I.V. stat. and repeat at 10—20 minutes period according to the blood gases so that the PH is greater than 7.3. (d) Give propranolol (Inderal) 0.01 mg/kg I.V. stat repeat that doses in 5—10 minutes if no response. Watch for bradycardia or hypotension during Inderal administration. (e) If breathing is slower or the patient is gasping then intubate and ventilate with an Ambu bag. 7. If spell continues for more than 60 minutes and the state of consciousness is unimproved consider emergency surgery and anaesthesia with gases containing high mixture of oxygen. Arrange portable X-Ray chest film and E.C.G. continue Bicarbonate I.V. as 10% solution (Add 10 CC $NaHco_3$ in 100 CC 5DW/1/5 saline)prior to shunting surgery.

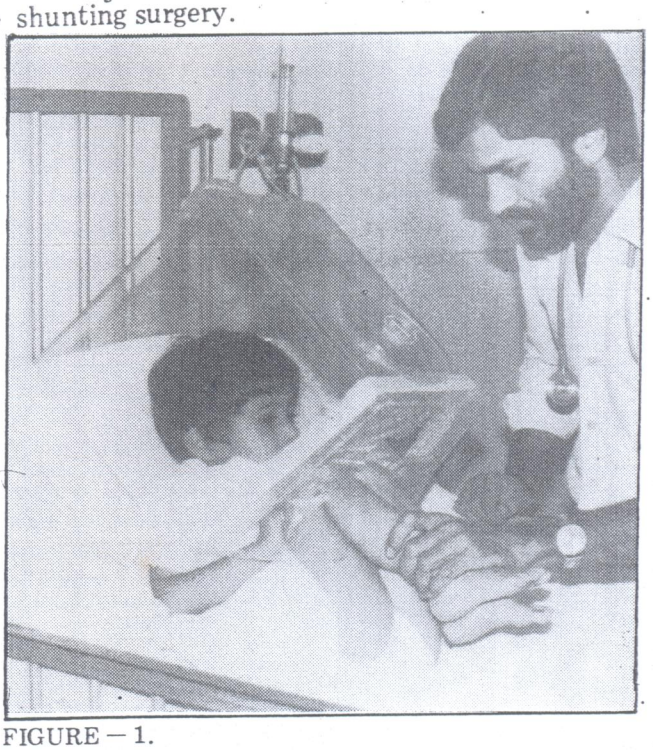

FIGURE — 1.

Photograph showing the method of controlling a cyanotic spell. Note the correct method of knee-chest positioning and oxygen inhalation.

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Note : DO NOT DO THE FOLLOWING :-

(a) Do not give diuretics such as Lasix (Frusamide) etc. (b) Do not give inotropic agents such as : (i) Digoxin, (ii) Isuprel or (iii) Adrenaline. (c) Do not give agents which would reduce blood pressure. (e) Do not treat or misdiagnose these spells as (i) Congestive cardiac failure, or, (ii) Broncho-Pneumonia.

Drug treatment of recurrent cyanotic spells:

These spells have a tendency to recur and Inderal (Propranolol) prevents recurrences. Start with 1–2 mg/Kg/day in two to four doses. Because of the availability of only tablet form of propranolol, in infants it may be practical to give two doses, although 4 daily doses are preferable. If spells are not completely controlled with a small dose the amount can be increased upto 3 mg/kg/day.

Inderal may not be useful in patients :

(1) Who are extremely cyanosed with Hemoglobin greater than 18 G%. (2) Or who have only a short grade 1–2 systolic murmur at the left sternal border. Note:– If the spells are controlled with Inderal continue the drug till the time of elective surgery. Sudden stoppage may lead to severe and prolonged cyanotic spell which may prove fatal.

In these patients shunting type surgical operation should be advised on urgent basis. Ideally all children with spells should have corrective surgery. The present scheme is outlined in consideration of local factors and resources.