Diagnosis and Treatment of Pulmonary Vasoconstriction Following Palliative Procedures in Congenital Heart Disease

Tohalazol hydrochloride (Priscoline) has been used in infants to reverse pulmonary arteriolar constriction in such diverse conditions as persistent fetal circulation and hypoperfusion in hyaline membrane disease. Its use in postoperative care in infants and children, however, is not well known. In this issue of Chest (see page 274), Moodie et al report successful use of tolazoline in a four-month old infant with transposition of the great arteries who developed pulmonary vasoconstriction following a palliative operation to enlarge the interatrial communication and assure better interatrial mixing. The postoperative chest x-ray film and radionuclide perfusion scan were consistent with reduced pulmonary perfusion. Hyperventilation with 80 percent oxygen did not relieve the vasoconstriction or improve the arterial oxygenation. However, following intravenous administration of tolazoline and blood volume replacement, the PaO₂ gradually increased, and a second chest x-ray film and radionuclide scan showed a significant improvement in pulmonary perfusion.

A similar set of circumstances is frequently encountered in neonates with transposition of the great arteries undergoing palliation by atrial balloon septostomy. Usually, there is a marked immediate improvement in aortic oxygen saturation after a successful septostomy. Most newborns with transposition of the great arteries have a patent ductus arteriosus which increases the pulmonary blood flow and favors the left to right shunting. Perhaps related to the acutely improved PaO₂ following septostomy, the ductal closure is accelerated, the left atrial filling and left to right shunting are soon reduced, and consequently, the arterial oxygen saturation falls, sometimes to alarmingly low values. Under these circumstances, a second septostomy may be attempted, only to find an adequate interatrial communication and a low left atrial pressure. Administration of prostaglandins may be successful at this point, since re-establishing the ductal patency will increase the pulmonary blood flow. However, when the drug is discontinued, the PaO₂ often falls rapidly, and the magnitude of ductal flow becomes prostaglandin-dependent. Patients with this clinical picture may then be subjected to surgical creation of an atrial septal defect which carries a high mortality, in part because its function is the same as that of balloon septostomy which it supplants. The Mustard operation with obligatory rerouting of venous streams, however, should be successful in infants who remain very cyanotic in the presence of an adequate atrial septal communication.

All possible mechanisms responsible for inadequate arterial oxygen saturation after a technically successful atrial balloon septostomy have not yet been clarified. Low pulmonary blood flow due to delayed resolution of neonatal pulmonary vascular resistance may well be one of the factors, especially after rapid functional closure of the ductus and consequent drop in aortic oxygen saturation. Under such circumstances, administration of tolazoline may prove to be effective.
In one of our infants with transposition of the great arteries and intact ventricular septum, the first septostomy carried out on the first day of life improved the initial aortic oxygen saturation of 24 percent (in high FiO2) to 78 percent (in room air). A ductus arteriosus with bidirectional shunting was demonstrated. After a few days of stability, the infant rapidly deteriorated and underwent a second septostomy at eight days of age. The atrial septal defect felt adequate in size. The ductus was no longer patent, and the aortic oxygen saturation in high FiO2 was 39 percent. The pressure in the left atrium (a 4, v 7, m 4) was lower than in the right atrium. The pulmonary artery pressure was 32/12 mm Hg, and there was no left ventricular outflow stenosis. During the septostomy, the infant became severely hypoxic and acidotic. The oxygen saturation in the femoral artery was 24 percent, and in the pulmonary artery, 93 percent. After tolazoline, while still in high oxygen, the oxygen saturation rose to 38 percent in the femoral artery and fell to 86 percent in the pulmonary artery, indicating a more favorable atrial mixing presumably as a result of an improved pulmonary blood flow. The infant required supplemental oxygen for weeks. A radionuclide perfusion scan at four weeks of age was consistent with low pulmonary artery perfusion. However, the cyanosis gradually improved, and when catheterized at five months of age, the aortic saturation at room air was 64 percent. At the Mustard operation at six months of age, the atrial septal defect was measured 1.0 cm in diameter. At catheterization 18 months later, the pulmonary artery and left ventricular pressures were normal.

In the report of Moodie et al, the cause of postoperative pulmonary vasoconstriction was probably pulmonary hemorrhage. During the Blalock-Hanlon operation, the pulmonary veins are cross-clamped, and pulmonary hemorrhage can occur even with the pulmonary artery occluded because of the excessive bronchial collateral flow originating from the aorta.

We have used tolazoline successfully in infants and children also following intracardiac repair of transposition of the great arteries, complete AV canal, and anomalous pulmonary venous drainage, in whom pulmonary artery monitoring revealed persistence or development of pulmonary hypertension. It might also be wise to consider the use of tolazoline postoperatively after palliative surgery when there is evidence that pulmonary vasoconstriction may be present. There may be a need for a greater role of radionuclide perfusion studies in the postoperative patient with congenital heart disease, especially when pulmonary artery monitoring is not available.

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REFERENCES

Positive End-Expiratory Pressure, Constant Positive Airway Pressure, and Cardiac Performance

We were warned by early investigators that maintenance of positive pressure over a prolonged portion of the ventilatory cycle will predictably result in a fall in cardiac output.1,2 This hemodynamic effect is due primarily to restriction of venous return into the chest by positive pleural pressure and to restriction of left ventricular filling by the action of positive alveolar pressure on alveolar capillaries.3

Suter et al4 provided an important service to the critically ill in 1975, when they showed that cardiac output could be improved in certain patients by the addition of positive end-expiratory pressure (PEEP) during mechanical ventilation. Although this observation had been made in isolated instances before, most clinicians remained concerned about the deleterious effects of PEEP reported in the 1930s and 1940s.

The hemodynamic response to ventilation with PEEP depends on, among other factors, left ventricular filling; one can compensate for the decrease in venous return that accompanies the use of PEEP by loading with fluids.5 Very high levels of PEEP can now be used without significant impairment of hemodynamic function.6

We have come full-circle in our use of positive pressure in the management of critically ill patients. We no longer restrict PEEP from use in patients with failing circulatory function. The ubiquitous capability for hemodynamic monitoring enables us to carefully choose an appropriate ventilatory mode and pressure for each patient’s individual needs. In studies performed in my institution,7 the use of PEEP was shown to improve cardiac output in the majority of patients with shock following myocardial infarction when left ventricular filling, as measured by the pulmonary arterial wedge pressure, was greater than 18 mm Hg. This finding is consistent with the observation by other investigators8,9 that a