PROBLEM OF AORTIC VASCULAR RINGS AND OTHER ANOMALIES OF THE AORTIC ARCH

A Case Report

BY

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SUMMARY

Anomalies of the aortic arch are not rare. Usually the lesions are asymptomatic and require no therapy. When symptoms such as tracheal obstruction or dysphagia are present in infants, roentgenographic studies will establish the diagnosis. Surgery is directed at the relief of the compression of the trachea and/or oesophagus. Some of the problems involved in infants with anomalous vascular rings are illustrated in a case report. A combination of surgical mediastinal dissection, endotracheal anaesthesia, and previous long-standing compression of the trachea and/or bronchi with possible pressure atrophy, all combined to invite the postoperative respiratory difficulties noted.

Anomalies of the aortic arch are not rare and can assume many diverse forms. Anomalous vessels may exist without producing any symptoms and may never require treatment. When symptoms are present, surgical correction is usually directed at the relief of the compression of the trachea and/or oesophagus and not at the production of a normal vascular pattern.

Hommel, in 1737, was one of the earliest investigators to publish an illustration of a double aortic arch, while Bayford, in 1794, described the dysphagia secondary to oesophageal obstruction due to a retro-oesophageal subclavian artery. The clinical syndrome of a double aortic arch was described by Wolman in 1939, and the first successful surgical correction of this anomaly was achieved by Gross in 1945.

In order to obtain an understanding of anomalies of the aortic arch, it is necessary that the various forms be recognized as bearing some developmental relationship to each other. The most satisfactory orientation regarding the developmental relationship of these anomalies is one which utilizes the concept of a primitive basic pattern consisting of a double aortic arch with both a left and a right patent ductus arteriosus and a midline descending aorta (fig. 1). Normal development of the human aortic-arch system includes progressive development as well as regression (Benson et al., 1962). Asymmetry is another characteristic of normal development and the final normal aortic-arch system is one in which the arch passes anterior and to the left of the trachea and then proceeds as the descending aorta.

Occasionally, failure of regression of either fourth aortic arch results in a persistent double aortic arch. The ascending aorta bifurcates into two branches, one which passes anteriorly and to the left of the trachea, while the other progresses posteriorly and to the right of the oesophagus, both limbs then join to form the descending aorta. In most cases, the right posterior arch is the larger of the two. Therefore, the left anterior arch is the one which is usually surgically resected. In a few instances, a double aortic arch may appear in combination with a descending aorta which lies to the right of the vertebral column as was noted in our case.

The pre-operative diagnosis of aortic arch anomalies is usually made by radiology; however, localized persistent atelectasis or emphysema in the newborn should suggest the possibility of vascular compression of a major bronchus. Similarly there may be symptoms of tracheal or oesophageal compression which may include stridor, respiratory distress, repeated respiratory

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infections, and post-prandial vomiting or difficulty. Final evidence is furnished by plain anteroposterior and lateral films, plus tracheogram and barium swallow films (Neuhauser, 1946) which show constriction of trachea or oesophagus, usually at a level just above the carina.

The prognosis in these cases varies with the degree of tracheal obstruction, the nature of the anomaly and the treatment given. Infants who present with symptoms at an early age must be observed carefully, since tightness of the ring often increases with growth. Sudden death may occur. Lower respiratory tract infections are common, some pneumonic, most resembling severe laryngotracheobronchitis. After operative correction, one expects stridor to persist for several weeks even though dyspnoea has been relieved more promptly.

Successful surgical correction has been accomplished in all types of vascular ring anomalies. Operation is still hazardous, especially for double aortic arch. In selecting the proper time for operation, the risk must be balanced against the degree of respiratory difficulty. If this is severe from the start, surgery is indicated in the neonatal period. If possible, surgery is delayed but each respiratory infection must be treated early and vigorously.

Some of the problems involved in infants with anomalous vascular rings are illustrated in the following case.

CASE REPORT

The patient, a 4-month-old infant was admitted because of respiratory distress. This male child was born at term after a normal pregnancy and delivery, weighing 3.6 kg. The infant breathed spontaneously, was not cyanotic, was slightly pallid, but required no resuscitative therapy. From the first day postpartum, the mother noted that the child had a continuous stridor which decreased in intensity with sleep and with alteration of posture.

At 12 weeks of age, the child developed a respiratory infection with concomitant respiratory distress which was diagnosed as pneumonitis. A second admission occurred 3 weeks later for respiratory distress; however, on neither occasion was the child noted to be cyanotic. The infant was treated with antibiotics, a highly moisturized, oxygenated atmosphere, and an oral iron preparation. The diagnosis of a vascular ring constricting the trachea and right bronchus was made on the basis of an oesophagram (fig. 2).

The infant on admission was well developed, well nourished (6.8 kg), somewhat pallid but without any cyanosis or clubbing. There was a continuous crowing stridor which decreased in intensity when the child was able to hyperextend the head and neck. Crying was associated with mild intercostal and abdominal retraction. Vesicular breath sounds were heard throughout the chest and no moist rales were heard. The heart was normal in size and no murmurs were heard. The pulse rate was 120 beats/min and the arterial pressure was 100/70 mm Hg. The child’s feeding appeared normal and vomiting was never noted. The liver edge was palpable. Oesophagram revealed a right aortic arch and a defect on the barium-filled oesophagus to the right and posterior margin of the oesophagus. Some narrowing of the trachea was also noted at this level.

Laboratory data were as follows: haemoglobin 10 g/100 ml, venous hematocrit 33 per cent, and total leucocyte count 11,000/cu.mm with a normal differential count. Urine and serum electrolytes were within normal limits. The electrocardiogram was interpreted as normal.
Four days later operation was performed. Premedication consisted of atropine 0.2 mg administered intramuscularly 1 hour before induction. The pre-operative pulse rate was 140 beats/min and the arterial pressure 110/70 mm Hg. Anaesthesia was induced with halothane, nitrous oxide and oxygen mixture using a semi-closed system utilizing a Heidbrink infant circle system. Intubation with a 3.5 mm Portex orotracheal tube was accomplished with 20 mg suxamethonium administered intravenously. A nasogastric tube was passed after induction to permit deflation of any gaseous dilatation of the stomach which might have impeded both respiration and surgery. The patient was placed in the right oblique position on a warm water blanket to maintain normothermia, and monitored electrocardiographically. Anaesthesia was maintained with nitrous oxide 2 l/min and oxygen 2 l/min with 0.5 per cent halothane added via a Fluotec vaporizer outside the circuit. Ventilation was manually controlled throughout the entire procedure.

An anterolateral thoracotomy in the third intercostal space was made and left pleura entered after division of the third and fourth costal cartilages. After excision of a large part of the left lobe of the thymus and retraction of the left lung, exploration of the mediastinal contents revealed a double aortic arch with right descent. The ligamentum arteriosum was transfixed and divided. The left or anterior aortic arch was then clamped and cut at its junction with the posterior arch where the descending aorta began, posterior to the origin of the left subclavian artery. The ascending aorta was elevated with sutures to the sternum (fig. 3).

![Diagram](https://academic.oup.com/bja/article-abstract/41/10/898/252730/0)
The left lung was expanded fully, a tube placed in the left pleural space and chest closure completed. During the procedure the patient was given 100 ml of whole blood (to replace the calculated blood loss). A marked improvement: pH 7.26, Pco₂ 51 mm Hg, oxygen saturation 95.7 per cent. Approximately 33 hours post-operative, the patient developed ventricular fibrillation which responded to intravenous phenytoin and oxygen saturation 95.2 per cent.

Satisfactory ventilation was achieved with this ventilator. The arterial blood-gas values showed evidence of marked improvement: pH 7.26, Pco₂ 51 mm Hg, and Po₂ 315 mm Hg.

The inspired oxygen concentration was slowly reduced to 40 per cent and the infant thereafter manifested improved blood-gas findings: pH 7.36, Pco₂ 53 mm Hg, Po₂ 120 mm Hg, haematocrit 32 per cent, and oxygen saturation 95.7 per cent. Approximately 12 hours later the child developed generalized epileptiform seizures which responded to intravenous phenobarbitone 240 mg. There was no evidence of increased intracranial pressure, serum electrolytes were normal, arterial blood-gas values were essentially normal: pH 7.44, Pco₂ 26 mm Hg, Po₂ 167 mm Hg, haematocrit 30 per cent and oxygen saturation 95.2 per cent.

As the hours progressed, it became more and more difficult to control ventilation because tenacious secretions intermittently plugged the tracheobronchial tree allowing the lungs to be ventilated only intermittently. The position of the distal end of the endotracheal tube was verified radiologically to ensure that it had not entered the area of tracheal stenosis or into either main bronchus. Tracheostomy was not performed at this time because it was believed that controlled ventilation and suction to the trachea could be accomplished through an endotracheal tube.

Since the patient's condition continued to deteriorate, antibiotics (aqueous penicillin 100,000 i.u. 6-hourly with streptomycin 160 mg 12-hourly) and steroids (hydrocortisone 25 mg 6-hourly) were added to the therapeutic regime. Approximately 33 hours post-operatively, the patient developed ventricular fibrillation but was successfully resuscitated by means of an external electrical defibrillatory shock and external cardiac massage with return of normal sinus rhythm. Within 30 minutes the infant developed cardiac arrest and subsequently died despite vigorous resuscitative measures. The blood-gas studies at a point prior to the cardiac arrest revealed: pH 7.11, Pco₂ 53 mm Hg, Po₂ 101 mm Hg, haematocrit 30 per cent, and oxygen saturation 87 per cent. During the entire 33-hour post-operative period, the patient's intravenous intake consisted of whole blood 45 ml, 5 per cent dextrose in water (to replace the calculated blood loss), 902 ml of NaCl 0.9 per cent, KCl 25 m.equiv/1., MgCl₂ 5 m.equiv/1., HPO₄⁻/ 3 m.equiv/1., and HCO₃⁻ 23 m.equiv/1.

At autopsy, the main findings were pulmonary oedema, anasarca, and bilateral pleural effusions. The tracheobronchial tree contained large amounts of mucoid, frothy material. The trachea at a level approximately 0.5 cm above the carina was compressed anteroposteriorly as was the right main stem bronchus close to the carinal bifurcation. The right main stem bronchial cartilages were incomplete and gave the appearance of congenital bronchomalacia. The oesophagus at the level of the carina was also narrowed by the encircling vascular ring formed by the double aortic arch with right descent and the fibrous ligamentum arteriosum. Although both ventricles showed a moderate dilatation, the cardiac valves, coronary vessels, and non-patent foramen ovale all appeared normal.

**DISCUSSION**

This case demonstrates a series of problems common to patients undergoing major cardiac surgery. Postoperatively, there is need for increased ventilation in order to maintain an adequate oxygen tension which thus avoids cardiac arrhythmias and satisfies the demands of an altered metabolism. The work of breathing is significantly increased, and the patient who cannot comply with this increased load develops an increasing hypoxic state.

Our patient at the conclusion of the procedure showed signs of depressed respiration necessitating prolonged intubation in order to clear the airway of secretions and to administer assisted ventilation via a mechanical ventilator. The choice of an automatic ventilator suitable for this child was extremely small in spite of the compensatory modifications that can be made to these ventilators making them applicable for paediatric use.

Although some clinicians note that the volume-variable ventilators such as the Bird Mark 8 or the Bennett PR2 appear preferable in the paediatric age group compared with the volume-limited type such as the Engström ventilator, our patient did not respond to this therapy as shown by clinical evaluation and by serial determinations of arterial oxygen and carbon dioxide tensions.

The Engström ventilator ameliorated the gas tensions in our patient, but his clinical status continued to deteriorate rapidly. In retrospect, one wonders whether a Bennett PR2 with a Q circle fitted with an expiratory retard mechanism may have proved more effective. Since deter-
minimations of postoperative respiratory volumes had not been made, a servo spirometer with an accuracy of 2 ml might have allowed a more precise detection of abnormalities if one had been available.

Other than postoperative physiotherapy utilizing an intermittent positive pressure breathing apparatus, treatment was directed towards: temperature control as gauged by a thermistor and utilizing an automatic temperature controlled water-alcohol blanket; constant electrocardiographic control; humidification of the atmosphere; cautious use of antibiotics; and continuous measurement of central venous pressure, peripheral arterial pressure and pulse.

It would have been most useful to have had the additional use of a thermistor cardiac output measuring apparatus, a means of assessing standard bicarbonate values as well as plasma electrolytes, a central arterial pressure recorder, an hourly recording of not only urine volume but also its specific gravity to aid in the regulation of fluid intake, and finally, the possible use of a hyperbaric chamber which has been most useful in congenital cardiac lesions.

One must conclude from this case that constant clinical evaluation of the paediatric cardiac patient pre-operatively, operatively, and post-operatively, is insufficient in maintaining an adequate cardiorespiratory status. Decisions concerning tracheostomy versus prolonged endotracheal intubation, the IPPB apparatus of choice, the regulation of electrolytes and water metabolism, and significance of laboratory parameters must be weighed rapidly and conclusively. When such a regime is not instituted, the patient’s increased work of breathing may produce an increased hypoxic state, respiratory acidosis, and congestive failure as noted in our patient.

REFERENCES