

## DOUBLE AORTIC ARCH\*

BY

R. E. STEEN AND S. J. DOUGLAS

*From the National Children's Hospital, Dublin*

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"Aortic ring," "bifid aortic arch," and "split aortic arch" are names given to a congenital malformation where, as a result of persistence of both fourth branchial arches (Fig. 1), the aorta, instead of being a single tube, divides into two, one branch going in front of the trachea, the other behind and almost invariably behind the oesophagus as well, the two limbs uniting again to form one descending aorta. The trachea and oesophagus are therefore gripped as it were in a vice between the two limbs, and it is easy to see how urgent obstructive symptoms may easily arise from what would otherwise be quite a trivial upper respiratory infection (Fig. 2). The posterior limb of the arch, spoken of as a retro-oesophageal aorta, usually carries most of the blood, the anterior limb being practically invariably the smaller of the two.

Recognition of vascular anomalies in this region goes back as far as 1794, when Bayford described dysphagia lusoria from the Latin *lusus naturae*, meaning a "freak of nature." The dysphagia, due to this particular freak, was in this case the result of an anomalous right subclavian artery, taking origin from the left side and passing over behind the oesophagus to the right side of the body. This is still the commonest anomaly according to Neuhauser, of Boston, but it is usually symptomless. In recent times Bedford and Parkinson (1936) have shown that a retro-oesophageal aorta may occur in two other conditions besides a double aortic arch. First, where in association with a right aortic arch, that is one which arches to the right side instead of the left, there is a ductus arteriosus originating in the normal manner from the sixth left branchial arch. This drags the descending aorta behind the oesophagus and down on the left side. Secondly, a similar state of affairs can occur with a left subclavian artery originating from a right aortic arch and travelling across from right to left, dragging the aorta over in the same way. Indeed, Paul (1948)

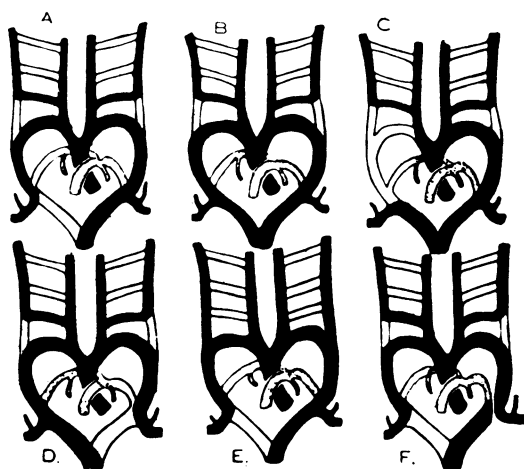
has described an even rarer form of retro-oesophageal aorta, namely one in which the aortic arch is on the left side in the normal manner, but where the descending aorta is dragged across from left to right by an aberrant right subclavian artery. (Bayford's original case and similar ones are due to a right subclavian artery alone passing behind the oesophagus, whereas in Paul's case the aorta itself is dragged to the right and becomes retro-oesophageal.)

Helen Taussig (1947) holds that it is not possible to differentiate a retro-oesophageal aorta from any one of these causes from one due to a double aortic arch, but Gross, of Boston, would not agree with this, and our experience accords with his that at least in some cases it is possible to make the diagnosis with certainty.

Arkin (1926) was the first to describe a double aortic arch. Ten years later he had collected another five cases, all being in adults and for the most part symptomless. Apparently once the child reaches 2 or 3 years old the danger period is past and symptoms tend to disappear. Possibly some factor like growth causes superimposed respiratory infection to be less liable to cause obstruction. After this age a double aortic arch is so symptomless that, although in an adult the retro-oesophageal aorta still remains tightly packed between the oesophagus and the vertebral column, one may almost guarantee its benign nature, provided the individual does not get an aneurysm.

The recognition of a double aortic arch still remained largely academic, however, until very recent times, when interest has been stimulated in these vascular anomalies by advances in thoracic surgery. These have culminated in the work of Gross, of Boston, who, in his *Surgery of Infancy and Childhood* (1953), gives an account of 21 cases of double aortic arch where from the previous history death would almost certainly have occurred, and in which, as a result of his severing the vascular ring, complete relief of symptoms resulted in 16 cases.

\*Part of a paper read at the meeting of the Thoracic Society in Dublin in June, 1951, the subject for discussion being "Tracheobronchitis."



[Courtesy of Hamilton, Boyd, and Mossman]

FIG. 1.—Abnormalities in the development of the branchial arch arterial pattern. The persisting systemic arteries are represented in solid black; the sections in outline represent embryonic structures which have atrophied and disappeared; the pulmonary trunk, its branches, and the ductus arteriosus are stippled. (A) Normal pattern; (B) double aortic arch; (C) abnormal origin of the right subclavian artery; (D) right aortic arch; (E) absence of both common carotid arteries; (F) extreme degree of coarctation of the aorta, the descending aorta being supplied by the ductus arteriosus alone.

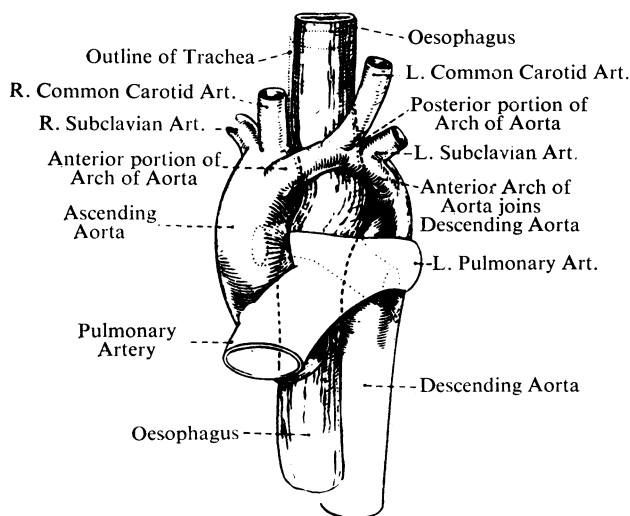


FIG. 2.—Double aortic arch. The diagram shows the aorta dividing into a narrower anterior branch which passes in front of the trachea, and a larger posterior branch passing behind the oesophagus.

#### SYMPTOMATOLOGY

This is as follows: (1) Stridor, often accompanied by a brassy cough due to pressure on the left recurrent laryngeal nerve; (2) dysphagia;

(3) attacks of severe acute obstructive laryngotracheitis due to a superimposed respiratory infection which would otherwise usually be a comparatively trivial illness.

#### DIAGNOSIS

The diagnosis of a retro-oesophageal aorta is a simple matter once the condition is suspected since, as Taussig points out, a teaspoonful of barium will clearly demonstrate the forward displacement of the oesophagus. The confirmation of a double aortic arch is then made by demonstrating as in our case that there was also pressure from in front (Figs. 3, 4, 5, and 6).

#### PROGNOSIS

This is a very uncertain matter during the first few years of life, as death may easily occur from asphyxia during feeding, or from an attack of acute obstructive laryngotracheitis with its complications.

#### TREATMENT

In Gross's cases he severed the narrower anterior limb and as a result completely relieved all symptoms. The only alternative line of treatment is palliative one to try to tide the child over until spontaneous recovery occurs at 2 or 3 years old.

#### CASE REPORT

Nuala, aged 7 weeks, referred to me by Dr. Courtney, of Nenagh, was admitted to the National Children's Hospital, Dublin, on February 25, 1944, with a history of stridor since birth.

She was an only child, and there was no history of rubella or other virus infection during the mother's pregnancy.

At first the stridor was only present when the child was receiving feeds, but during the fortnight before admission it had been constantly present and associated with a cough. A few days before admission the child's condition had deteriorated. On examination she was a poor colour with marked stridor, and the extraordinary muscles of respiration were in full action with inspiratory recession of the ribs. Sulphonamide, penicillin, and streptomycin were given together, and after a week what must have been superimposed bronchitis or bronchopneumonia subsided. The child still had stridor and marked asphyxial attacks which occurred always at feeding time and often lasted as long as five minutes, by which time on many occasions the child appeared dead. Thinking the condition might be a trachea-oesophageal fistula in which case the giving of barium always results in aspiration into the lungs and a fatal bronchopneumonia, 1 or 2 ml. "lipiodol" was introduced into the oesophagus. A diagnosis of double aortic arch was made. The question of operation was then considered and angiocardiology was discussed. I would have

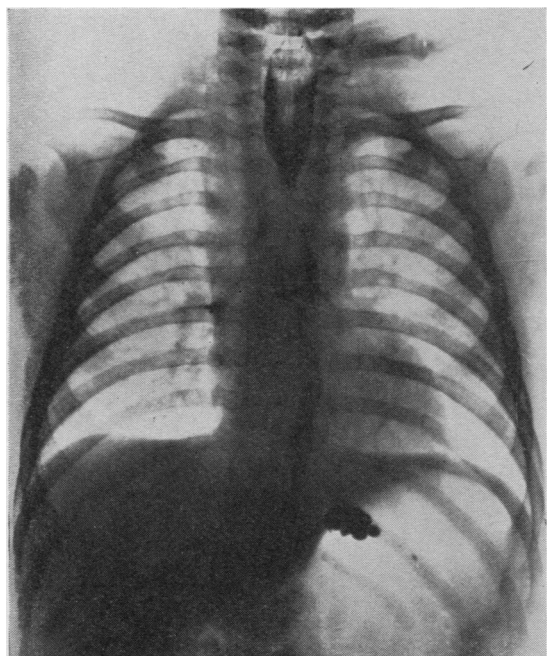


FIG. 3.—Postero-anterior view of the oesophagus outlined by a "lipiodol" swallow which shows the hold-up of "lipiodol" and double indentation of the wall of the oesophagus by a constricting vascular ring.

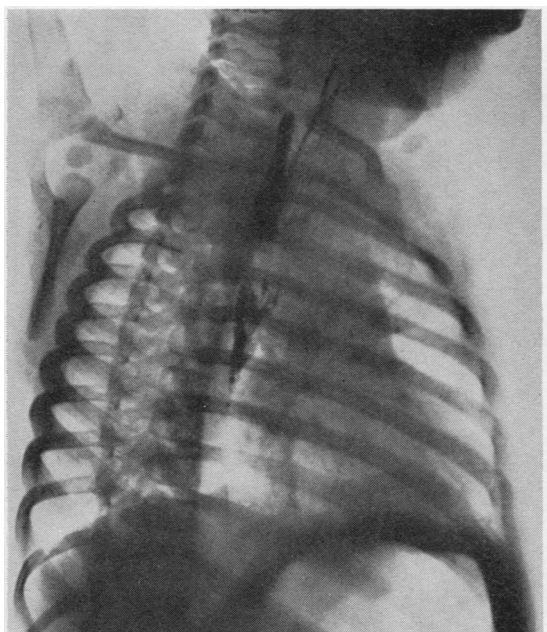


FIG. 5.—Right oblique view showing the hold-up in the oesophagus and narrowing of the trachea in front following "lipiodol" instillation into the trachea.



FIG. 4.—Lateral view showing the oesophagus pushed forward and the narrowed tracheal air-column in front and slightly higher.

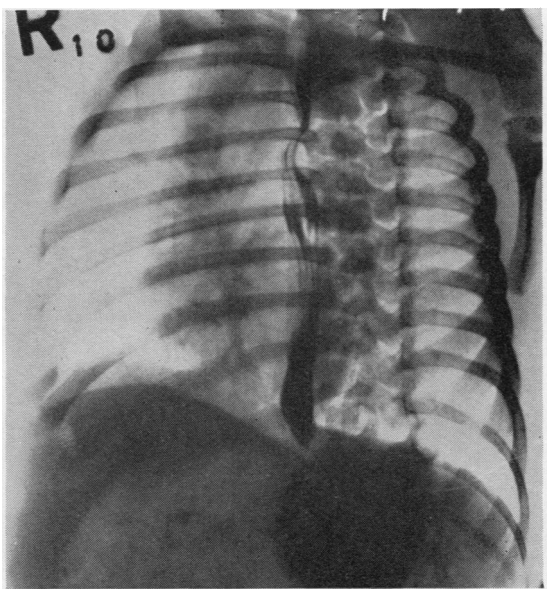


FIG. 6.—Left oblique view showing the barium-filled oesophagus pushed forward by the posterior branch (retro-oesophageal aorta) and just above this the indentation from in front of the anterior branch.



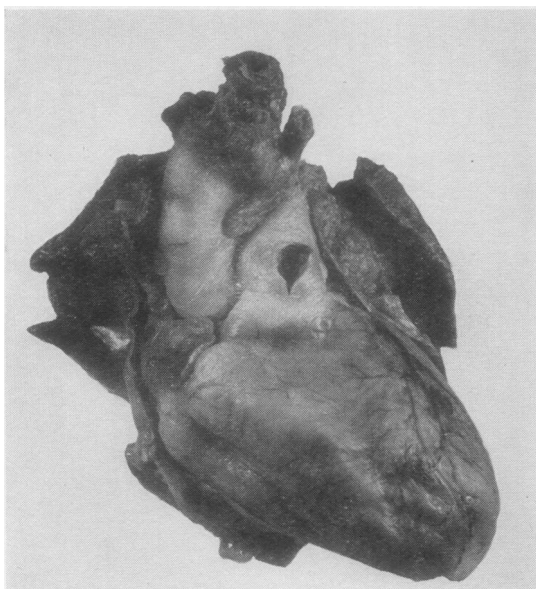


FIG. 7.—Anterior view showing the aorta dividing into the anterior and posterior limbs surrounding the trachea and oesophagus; pulmonary artery with vertical slit.

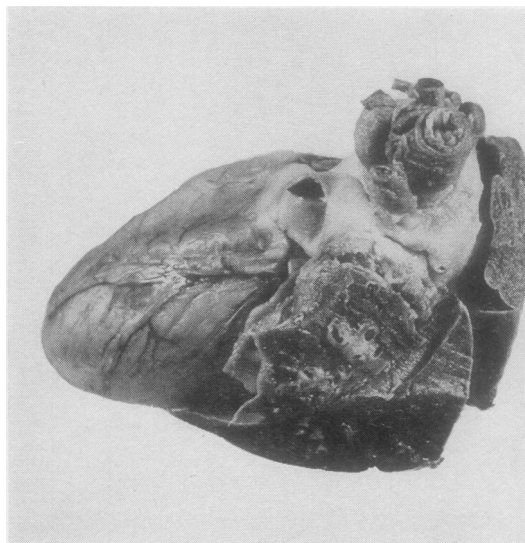


FIG. 9.—View taken from above to show the trachea and the oesophagus as it were caught in a vice between the two limbs of the aorta.



FIG. 8.—Posterior view showing the retro-oesophageal aorta.

liked very much to have done an angiogram, but we did not consider it justifiable when the diagnosis was unequivocal, since there is a definite slight mortality rate from angiography even in babies, and especially if the case was not going to be operated upon. By now the child was putting on weight at the normal rate and the asphyxial attacks at feeding time, which had been very numerous every day at the beginning, had now become infrequent at fortnightly or three-weekly intervals, although feeding had still to be very careful. One hoped also that superimposed infections might be controlled by sulphonamides and antibiotics, and it was finally decided to wait in the hope of a spontaneous recovery rather than risk an operation. We seemed to be making good progress when quite suddenly, towards the end of September, 1949, at the age of 9 months, and when she had reached a weight of 19 lb., she died from asphyxia during feeding.

At necropsy a double aortic arch was found with the larger limb passing behind the oesophagus and the narrower anterior limb passing in front of the trachea (Figs. 7, 8, and 9). The heart was otherwise normal.

It is easy to be wise after the event, but I think the decision to operate or not was a very close one. Mr. Henry and I discussed this case on innumerable occasions. Of course if there were no tendency to spontaneous recovery there would be no question that an operation must be carried out, but a number of cases recover in this manner.

The two main dangers appear to be asphyxia on the one hand, always associated with feeding, or superimposed infection leading to acute obstructive laryngotracheitis. The chance of fatalities from the latter are, I think, greatly minimized by antibiotics, and indeed I think this child would not have died from this cause, but even with careful feeding the risk of asphyxia is apparently very great. This baby very nearly came through on conservative treatment, and in mild cases with minimal symptoms this line of therapy appears to be the best plan, as surgical treatment carries a definite operative mortality. On the other hand, where symptoms, as in our case, were severe, and especially where asphyxial attacks, as in this child, were a feature, we should on another occasion feel inclined to give the verdict in favour of operation.

## SUMMARY

A case of double aortic arch is described. It is probable that this condition is more common than would appear from the number of recorded cases, and that its recognition and treatment would mean a saving of life.

We are indebted to Dr. Helen Taussig and the Commonwealth Fund for permission to reproduce Figs. 1 and 2.

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