Rare Anomaly of the Aortic Arch Combined
With Coarctation of the Aorta

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Introduction
Several reports on anomalies of the aortic arch concomitant with coarctation of the aorta are to be found in the literature. This paper describes such anomalies as they were found in two of our surgical cases, which showed unusual features not hitherto described in the literature available to us. These anomalies, although rare, are of practical importance with a view to differential diagnosis.

The etiology of coarctation is still obscure. The two cases presented in this paper should be taken into account in any attempt to explain the condition on the basis of embryological considerations.

The descriptions so far given in the literature stated that the right subclavian artery or the right aortic arch occurred distal to the coarctation. We were unable to find a single report on a right subclavian artery or a double arch proximal to the coarctation. And it was precisely this condition which we found in the two patients to be described.

Edwards recently described a case of Taussig-Bing complex with coarctation, in which the right subclavian artery arose proximal to the coarctation.

In the first of our two cases the right subclavian artery arose from the aorta above the coarctation; in the second case the double aortic arch was localized above the coarctation. In both cases the coarctation was found opposite the insertion of the ligamentum arteriosum.

Case 1:** M. V., female, aged 19. The onset of symptoms, in 1946, was characterized by pain in the chest, loss of weight and fatigueability, which continued until a severe hemoptysis occurred in 1946. Investigation at the time failed to reveal any tuberculous process but a mediastinal tumor was demonstrable by X-rays. The patient was referred to us at the St. Antonius Ziekenhuis, Utrecht.

The patient gave no impression of being very ill. The blood pressure determined in the arms was 150/90. Heart: slight systolic murmur. Pirquet tuberculin test: negative. Hematology normal. Fluoroscopy showed a tumor dorsal to the left main bronchus. The esophagus was displaced to the right. Tomography of the hilar region showed marked stenosis of the left main bronchus caused by the tumor. The presence of severe stenosis was confirmed by bronchography. Bronchosopic examination showed that the bronchial wall was normal and that the stenotic part was still patent.

Angiocardiography: apparently satisfactory filling of the tumor with contrast medium suggested a probable aneurysm or a highly vascular neoplasm. Another severe hemoptysis occurred during the patient's stay in the hospital, this time causing what was considered vital danger. The blood pressure of 150/90 did not strike us as hypertensive, and the possibility of coarctation was consequently not considered. An operation was performed in spite of an uncertain diagnosis.

The thorax was opened (March 3, 1949; surgeon: A. G. B.), and numerous adhesions were separated without difficulty. The first striking feature was found in the fact that a right subclavian artery arose distal to the left subclavian artery. The right radial pulse was suppressed when this vessel was clamped. The artery extended

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**This case included in the thesis of A. G. Brom; "De Coarctatio aortae en haar behandeling."
to the right, behind the esophagus. About 1.5 cm. distal to the division into the right subclavian artery a multicolored tumor the size of an orange was found to arise from the aorta, which it completely surrounded over a length of almost 6 cm. with the exception of the left side of the aorta; this side was free but showed a slight depression in the aortic wall. This tumor was an aneurysm of the descending aorta arising immediately distal to a constriction of the aortic lumen at the level of the insertion of the ligamentum arteriosum (Fig. 1, a, c). The lumen of the aortic arch was slightly wider than that of the subclavian arteries, both of which were of small caliber. The caliber of the descending aorta distal to the aneurysm exceeded that of the arch and that of the descending aorta proximal to the aneurysm. The latter part showed a ventral adhesion with the lung to the right of the median line and a dorsomedial adhesion with the esophagus. The aneurysm was separated from the lung. At this point a hemorrhage from the aneurysm occurred, which was controlled with a finger. Dissection of the aorta was then attempted. This dissection and the subsequent ligation of the intercostal vessels were accomplished by routine procedure, in the course of which part of the membranous portion of the left main bronchus was resected with the aneurysm. The defect was closed with a free pleural graft.

The presumption that the aneurysm was the cause of the stenosis of the main bronchus and of the hemoptysis was confirmed. The minimal length of aorta to be removed in view of the stenosis and the aneurysm left a defect of 6 cm. End-to-end suture of the aorta was impossible. Since no aortic graft was available at the time (1949) the anomalous right subclavian artery was used to bridge the defect; an end-to-end anastomosis with the distal part of the aorta was made. This anastomosis provided a newly made aorta of lesser caliber than the arch but with a circumference exceeding 4 cm. (Fig. 1, b).

The operation was completed by routine procedure.

Pathology

Macroscopy: The specimen had the size of an orange and consisted chiefly of a blood clot. A transverse section showed intermittent, probably aneurysmal dilations in the structure of the vascular wall. The vascular wall
seemed to be of the usual thickness but the wall of the aneurysm was thinner.

**Microscopy:** the internal layer of the wall of the aneurysm was completely hyaline, with split formations, and with an inner lining of fibrin. The external layer showed a lamellar arrangement of hyaline fibres, with several diffusely spread round cells. No muscular tissue was found, and elastic tissue was only found in small scattered quantities. The irregular, broad adventitia contained a larger quantity of elastic tissue. A transverse section of the aorta showed marked intermittent thickening of the intima caused by degenerative changes (split formations, lipid deposits, hyalinization, necrosis and mucoid changes). Some degree of round-cell infiltration was seen in the irregular adventitia. The media was broad, causing the basic constriction of the lumen, and remarkably thick elastic bundles were found in between, with occasional patches of hyaline fibrous and muscle tissue. There was a marked diminution of fibrous elements in favour of elastic structures.

The postoperative course was uneventful. No pulse was palpable in the right arm. The patient was discharged three weeks after the operation and several periodic follow-ups were subsequently made.

![Figure 2: X-ray of R. H. Marked left-sided enlargement of the heart. Esophagus showing impression of retro-esophageal vessel.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21292/ on 06/22/2017)
The last follow-up was made in March, 1954, i.e., five years after the operation, when the patient was referred to us by her obstetrician. She was pregnant and complained of headaches and dizziness. Nothing anomalous was found at examination. The pulsation of the femoral arteries synchronized with that of the left radial artery. The right radial artery was palpable but the amplitude was inconsiderable. Intra-arterial determination yielded blood pressure values of 120/80 in the left arm and 110/80 in the legs (Fig. 1, d, e).

***Case 2***: R. H., female, aged 2, of normal weight and with a deep, square thorax, slight scoliosis and atrophy of the limbs.

At the age of 21 months there was a period of left heart failure with unrest and marked orthopnea. Coarctation with pre- and post-stenotic hypertension was diagnosed. The blood pressure reading was 200/130 in the arms and 150/110 in the legs. Pulsation of collateral arteries was felt in both axillary regions. The ECG showed a sinus tachycardia of 144 with left axis deviation and left ventricular preponderance. Fluoroscopy revealed marked cardiomegaly, with presumable predominance of left-sided configuration; there was no evidence of pulmonary congestion. Contrast radiography of the esophagus did not indicate post-stenotic dilation of the aorta. The investigator at the time overlooked the constant impression of a retro-esophageal vessel extending in right superolateral direction such as is seen in cases of anomalous right subclavian artery or double aortic arch (Fig. 2). During angiocardiography the circulation was relatively slow, with satisfactory contrast in the right and left heart. The left auricle and ventricle were enlarged but the ascending aorta was not; there was circumscribed stenosis of normal localization. No post-stenotic dilation was observed.

Since there was no unmistakable evidence of left heart failure, no pre-operative digitalization was given.

During the operation (September, 1955; surgeon: A. G. B.) the patient had 16 attacks of cardiac failure with brief disappearances of blood pressure. Heart massage

![FIGURE 3: (a and c) Preoperative and postoperative findings and (b) photograph of the resected specimen.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21292/)
was required during these periods. Surgical procedure was considerably impeded by this necessity, which made it almost impossible to secure a free field for dissection. In the course of dissection it was established that, immediately beyond the left subclavian artery, a second and equally large vessel arose from the aortic arch, extending to the right behind the esophagus. There was no cessation of the right pulse when this vessel was clamped off. A double aortic arch was therefore presumed to exist. The coarctation was localized at the site of junction of the ligamentum arteriosum. Dissection of the coarctation required considerable effort (due partly to the degree of weakness of the heart). Pott's clamps were placed on the aorta. The stenotic part was resected and an end-to-end anastomosis was made, using interrupted sutures (Fig. 3). The anastomosis obtained after removal of the clamps was certainly as large as the arch. Cardiac arrest recurred after closure of the thorax, which was then re-opened for heart massage. The blood pressure subsequently returned.

The attacks of cardiac failure may have been due either to hypersensitivity of the left hilus to manipulation or to mild pulmonary edema (the lung felt somewhat spongy).

The ECG recording during cardiac failure showed (Fig. 4) slowing of the sinus rhythm with occasional ventricular arrest (skipped beat). The deceleration in rhythm was followed by an increase in frequency associated with merging of the P-top with the preceding T-top. These findings seemed to suggest that, even during maximal tachycardia, the underlying rhythm originated in the sinus node in the normal manner.

In the course of the operation there was an alarmingly progressive widening of the QRS-segment with a marked depression in the ST-wave. Yet even at this stage the arterial pressure was fairly constant at 160/85 (and at 140/85 after elimination of the coarctation).

During the operation there was a steady rise in the temperature, which reached 41° C. at the end. The patient was cooled in an ice bath after the operation. She was removed from the bath at 36.7° C, but cooling continued. The ECG still showed marked widening

![ECG recordings](https://journal.publications.chestnet.org/pdffaccess.ashx?url=/data/journals/chest/21292/)
Discussion

A series of 172 cases of coarctation given surgical treatment since 1948 included the two above described cases of a rare anomaly of the aortic arch concomitant with coarctation of the aorta.

The first patient presented an anomalous right subclavian artery arising between the left subclavian artery and the coarctation. This vessel was undoubtedly the right subclavian artery, as shown by the fact that the pulse in the right arm disappeared as soon as the vessel was severed, whereas preoperative determinations revealed the same brachial pressure bilaterally. The pulse did not re-appear until three months after the operation; its amplitude was then inconsiderable. Follow-ups covering a period of more than five years showed, moreover, that the surgical results were in no way

![Image](image.png)

**FIGURE 5:** ECG limb leads a, b and c during anesthesia (a) before thoracotomy (b) after closure of the thorax and immersion in ice water, temperature about 36° C., (c) ten minutes after b, temperature about 34° C. (d) Return to normal rhythm.
impaired by the fact that the anastomosis made was smaller in caliber than the proximal part of the aorta. The patient's blood pressure was on the low side rather than high, even during pregnancy. We disagree with Gross' as to his claim that the anastomosis should have a lumen at least as large as that of the aortic arch.

Optimal results in adults are ensured if the circumference of the anastomosis is 4 cm. or more. We believe also that the shape of the arch following anastomosis is of importance and should approximate the normal anatomical aspect as closely as possible.

In the second case a wide, patent right aortic arch was found proximal to the coarctation. Both arches were nearly equal in caliber, the right being slightly less wide than the left. Pre- and postoperative determinations revealed almost the same pressure in both arms. The pulse in the right arm remained unaltered throughout the operation, even when both arches were clamped. The preoperative esophageal findings were such as to suggest an aortic ring although there were no symptoms of dysphagia lusoria. The complete absence of tracheal compression and the unusual technical difficulties encountered prevented severance of the right arch.

It should be emphasized that a characteristic picture of coarctation of the aorta was found in both cases.

**SUMMARY**

Two cases of rare anomalies of the aortic arch combined with coarctation of the aorta are described.

In the first case the anomalous right subclavian artery arose proximal to the coarctation and was successfully used to bridge the defect resulting from resection of the coarctation.

The second case presented a patent right aortic arch, likewise proximal to the coarctation. Intervention in this case had to be limited to the coarctation in view of the absence of tracheal compression and the technical difficulties encountered.

In both cases the aortic stricture presented the characteristic features of coarctation of the aorta.

**RESUMEN**

Se relatan dos casos de anomalías raras del arco aórtico combinadas con coartación del istmo.

En el primer caso la arteria subclavia anómala salía arriba de coartación y fué usada con éxito como un puente sobre el defecto después de la reseción de la coartación.

En el segundo caso arco aórtico derecho abierto se encontró otra vez arriba de la coartación. En este, tratamos sólo la coartación ya que no había compresión de esófago y también por las dificultades técnicas durante la operación.

En ambos casos el estrechamiento aórtico tenía las características de la estenosis ístmica de la aorta.
RESUME

Les auteurs rapportent deux cas d’anomalies rares de l’arc aortique associées à un rétrécissement de l’isthme.

Dans le premier cas une artère sous-clavière droite anormale prenait naissance au-dessus de la coarctation et put être utilisée avec succès pour rétablir une situation normale après résection de la coarctation.

Dans le second cas, un arc aortique droit évident fut constaté au-dessus de la coarctation. Dans ce cas, les auteurs ne se sont occupés que de traiter la coarctation, étant donné l’absence de compression oesophagienne, et les difficultés techniques apparues lors de l’intervention.

Dans les deux cas, le rétrécissement aortique présentait le tableau caractéristique des sténoses isthmiques de l’artère.

ZUSAMMENFASSUNG

Bericht über 2 Fälle seltener Anomalien des Aortenbogens in Verbindung mit Verengung des Isthmus.

Bei dem ersten Fall entsprang die abnormale rechte Arteria subclavia oberhalb der Verengung und diente mit Erfolg zur Überbrückung des Defektes nach Resektion der Verengung.

Bei dem 2. Fall wurde wieder oberhalb der Verengung ein offener rechter Aortenbogen gefunden. In diesem Fall befassten wir uns nur mit der Verengung wegen des Fehlens einer Speiseröhren-Kompression und der technischen Schwierigkeiten während des Vorgehens.

In beiden Fällen wies die Aortenstriktur das charakteristische Bild einer Isthmus-Stenose der Aorta auf.

REFERENCES