

Right aortic arch with aberrant left subclavian artery Report of two cases

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Summary. Two cases of adult asymptomatic right aortic arch with an aberrant subclavian artery are reported. They were discovered at time of coronary surgery. Preoperative coronary arteriography failed to demonstrate the anomalies. In one case, the right arch was suspected on chest x-ray and preoperative barium oesophagography. In one case, the proximal suture of one saphenous bypass graft was performed on the left common carotid artery. Right aortic arch is a malformation rarely discovered in adults. It generally produces no symptoms when not associated with cardiac disease.

Crosse aortique droite avec artère sous-clavière gauche aberrante : rapport de deux cas

Résumé. Deux cas de crosse aortique droite avec a. sous-clavière aberrante sont rapportés. Ils ont été

découverts fortuitement à l'occasion d'un pontage coronaire. La coronarographie préopératoire n'a pas montré la variation. Dans un cas, la dextro-position de l'aorte a été suspectée sur les radiographies du thorax et sur l'oesophagographie. Dans l'un des deux cas, le greffon veineux saphène a été implanté sur l'a. carotide primitive gauche. La dextro-position de l'aorte est une variation rarement découverte chez l'adulte. En général, elle n'engendre aucun symptôme quand elle n'est pas associée à d'autres malformations cardiaques.

Key-words : Ascendant aorta — Aortic arch — Coronary surgery — Congenital heart disease

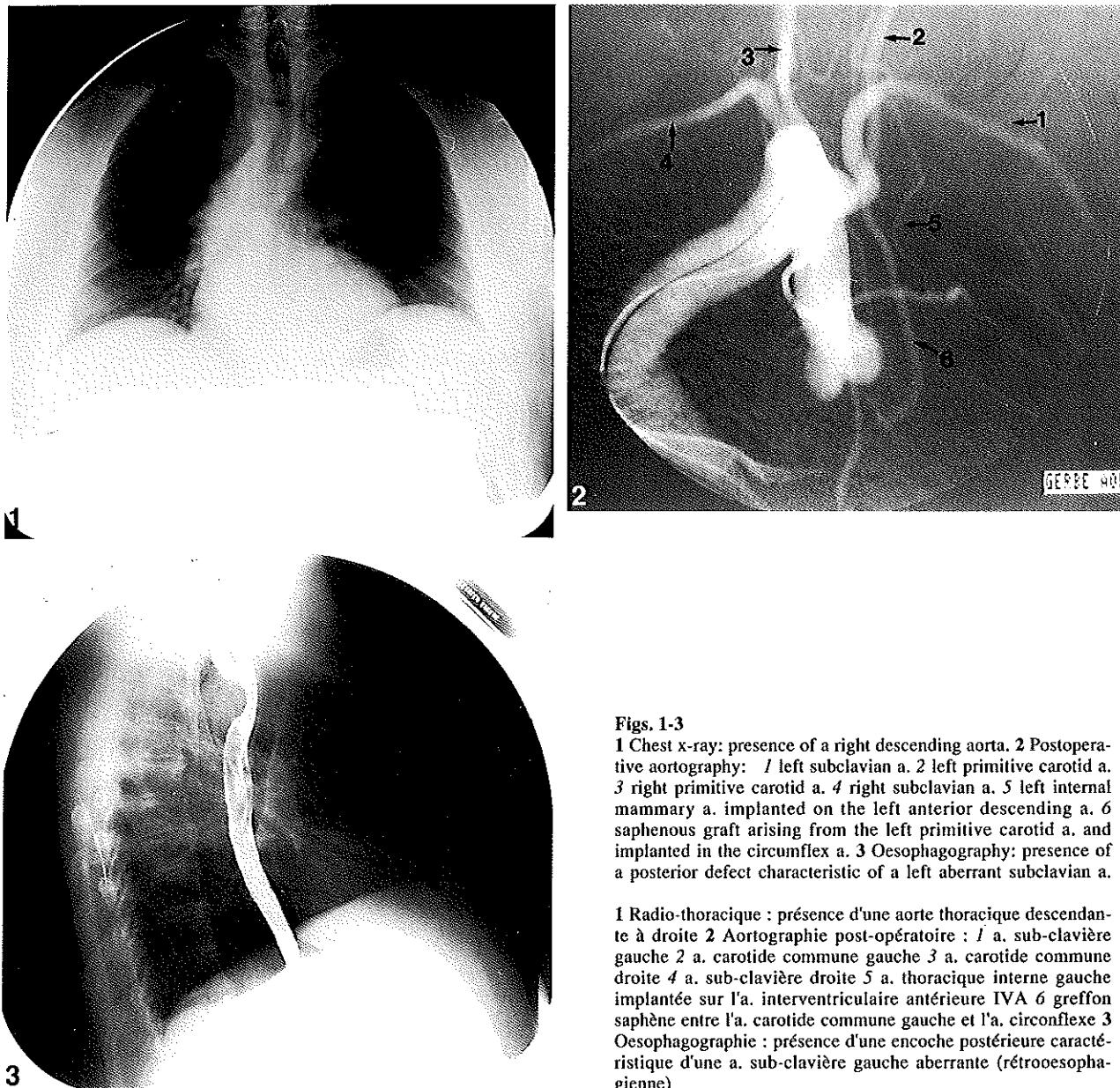
Right-sided aortic arch may be the part of a double aortic arch or the only channel at that level of the aorta. Its incidence is about 0.04 to 0.14% in adults [1, 6]. Whereas generally an incidental finding in the adult, symptomatic right arch is usually detected in infancy. Symptoms include tracheal obstruction,

cyanosis, and recurrent respiratory infection [5, 8, 9]. We report 2 adult cases of asymptomatic right aortic arch with aberrant left subclavian artery. They were encountered during 5,000 cardiac surgical procedures, representing 0.04% of incidence.

Case report

Case No 1

A 63 year old man underwent coronary artery bypass for severe multi-vessel disease. Except myocardial disease, past medical history was uneventful. In particular, neither dysphagia nor dyspnea were noted. On preoperative chest x-ray the presence of a right aortic knob and of a right descending aorta was missed by the radiologist (Fig. 1). At operation, after sternotomy and opening of the pericardial sac, a right-sided aorta was discovered. A supra-aortic vessel arising from the anterior aspect of the aorta crossed the anterior border of the trachea towards



Figs. 1-3

1 Chest x-ray: presence of a right descending aorta. 2 Postoperative aortography: 1 left subclavian a. 2 left primitive carotid a. 3 right primitive carotid a. 4 right subclavian a. 5 left internal mammary a. implanted on the left anterior descending a. 6 saphenous graft arising from the left primitive carotid a. and implanted in the circumflex a. 3 Oesophagography: presence of a posterior defect characteristic of a left aberrant subclavian a.

1 Radio-thoracique : présence d'une aorte thoracique descendante à droite 2 Aortographie post-opératoire : 1 a. sub-clavière gauche 2 a. carotide commune gauche 3 a. carotide commune droite 4 a. sub-clavière droite 5 a. thoracique interne gauche implantée sur l'a. interventriculaire antérieure IVA 6 greffon saphène entre l'a. carotide commune gauche et l'a. circonflexe 3 Oesophagographie : présence d'une encoche postérieure caractéristique d'une a. sub-clavière gauche aberrante (rétrooesophagiennne)

the left mediastinum. By mistake, this vessel was considered as the subclavian artery. Three saphenous grafts were implanted on five vessels: right coronary a., first diagonal branch (latero-lateral suture) and second diagonal branch, circumflex a. The left mammary a. was implanted in the left anterior descending a. Because of lack of place on the

ascending aorta, the proximal suture of the circumflex bypass was performed on the aberrant vessel. The postoperative arteriographic control (Fig. 2) showed patency of all bypasses with the mammary a. arising from an aberrant subclavian a. In fact, the circumflex graft was implanted on the left common carotid artery.

Case No 2

A 74 year old man underwent a triple coronary bypass. On the pre-operative chest x-ray the aortic knob was displaced to the right of the trachea and the oesophagus on the barium oesophagography showed a large indentation of its posterior wall (Fig. 3). At the time of sur-

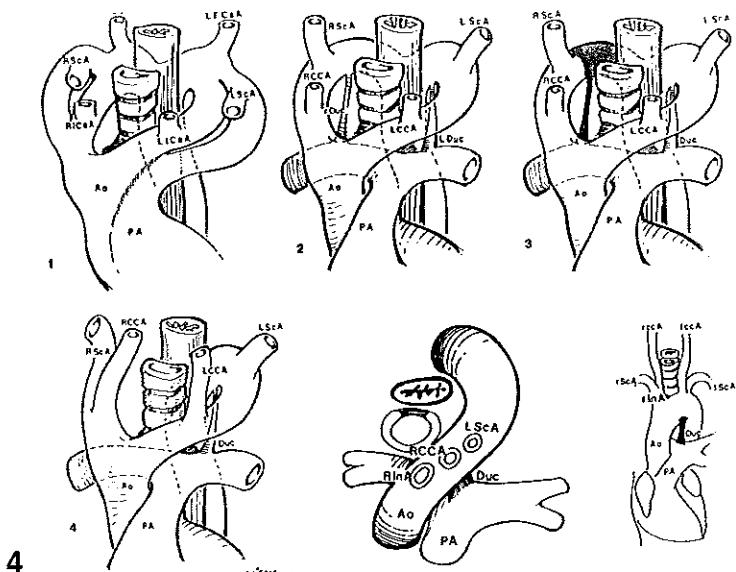


Fig. 4

Embryologic development of the left aortic arch. Normal regression occurs after subclavian a. Ao aorta PA pulmonary a. LICA a. left internal carotid a. RICA a. right internal carotid a. LECa a. left external carotid a. RECa a. right external carotid a. LCCA a. left common carotid a. RCCA a. right common carotid a. LScA a. left subclavian a. RScA a. right subclavian a. Duc ductus

Développement embryologique normal de la crosse aortique gauche. La régression normale survient après l'a. sub-clavière gauche. Ao aorte PA tronc pulmonaire LICA a. carotide interne gauche RICA a. carotide interne droite LECa a. carotide externe gauche RECa a. carotide externe droite LCCA a. carotide commune gauche RCCA a. carotide commune droite LScA a. sub-clavière gauche RScA a. sub-clavière droite Duc canal artériel (de Botal)

ry, after sternotomy, the left common carotid a. was the first vessel arising from the anterior aspect of the ascending aorta, however somewhat higher than in case 1. The left common carotid a. crossed the anterior side of the trachea. In this case, all aortic anastomoses could be performed on the ascending aorta.

Discussion

Variations in embryologic development of the aortic arch explain the formation of a right aortic arch. Edward's hypothesis (Fig. 4) considered an initial ventral aorta that gives rise to two aortic arches on each side [3]. Each arch joins its homolateral dorsal aorta. A left and a right ductus arteriosus is present. After fusion of the left and the right dorsal aortae, the descending aorta is in the midline. The common carotid aa. originate from the left and fourth superior arches which include the aortic arch. The left and right sixth arches, caudal to the preceding, correspond to the pulmonary arch from which arise the left and right pulmonary arteries. On each side, the subclavian arises from the

dorsal aortae before their fusion. Later, they migrate on the cephalic arch. Ultimately the ventral aorta separates into ascending and pulmonary systems.

Regression of the right cephalic arch occurs after the right subclavian a. conducts to the normal aortic system. In contrast, interruption of the left arch at different levels explains the persistence of a right fourth aortic arch [1, 2, 12]. Different types are distinguished considering branching of supraaortic vessels, position of the ductus arteriosus, shift in position of dorsal aorta, and eventual association with cardiac disease [6] (Fig. 5).

As in our 2 cases, the right aortic arch with aberrant left subclavian a. [7, 10, 12] results from regression of the left arch between the left common carotid and left subclavian aa. (Fig. 5a). The left common carotid a. arises as the first aortic branch, followed from before backward by the right common carotid a. and the right subclavian a. The aberrant left subclavian a. is the fourth branch, originating from the left side of the upper descending aorta. Then it runs towards the left arm, passing behind the oesophagus, indenting its posterior wall. In some cases, the distal portion of the

left arch remains like an enlargement protruding to the left (diverticulum of Komerell). The right arch passes over the right main bronchus to the right of the trachea and oesophagus. The thoracic aorta generally descends on the right side (see case 1), but may in some instances be in the midline or on the left, after crossing behind the oesophagus (type circumflex right aa.). In the latter case, or in the case of Komerell's diverticulum, the posterior indentation of the oesophagus is much greater than the one produced by the subclavian a. In the lateral projection it causes anterior bowing of the upper portion of the oesophagus (see case 2).

Tracheal or oesophageal compression depends to a large extent on the location of the ligamentum arteriosum [4], that is generally situated on the left side connecting the aberrant left subclavian artery to the left pulmonary trunk. Thus a vascular ring is formed by the right arch to the right, the left subclavian a. posteriorly, the ligamentum to the left and the pulmonary arterial bifurcation anteriorly. If only a right ligamentum is present, a vascular ring is usually not formed.

The second type is the right aortic arch with mirror-image bran-

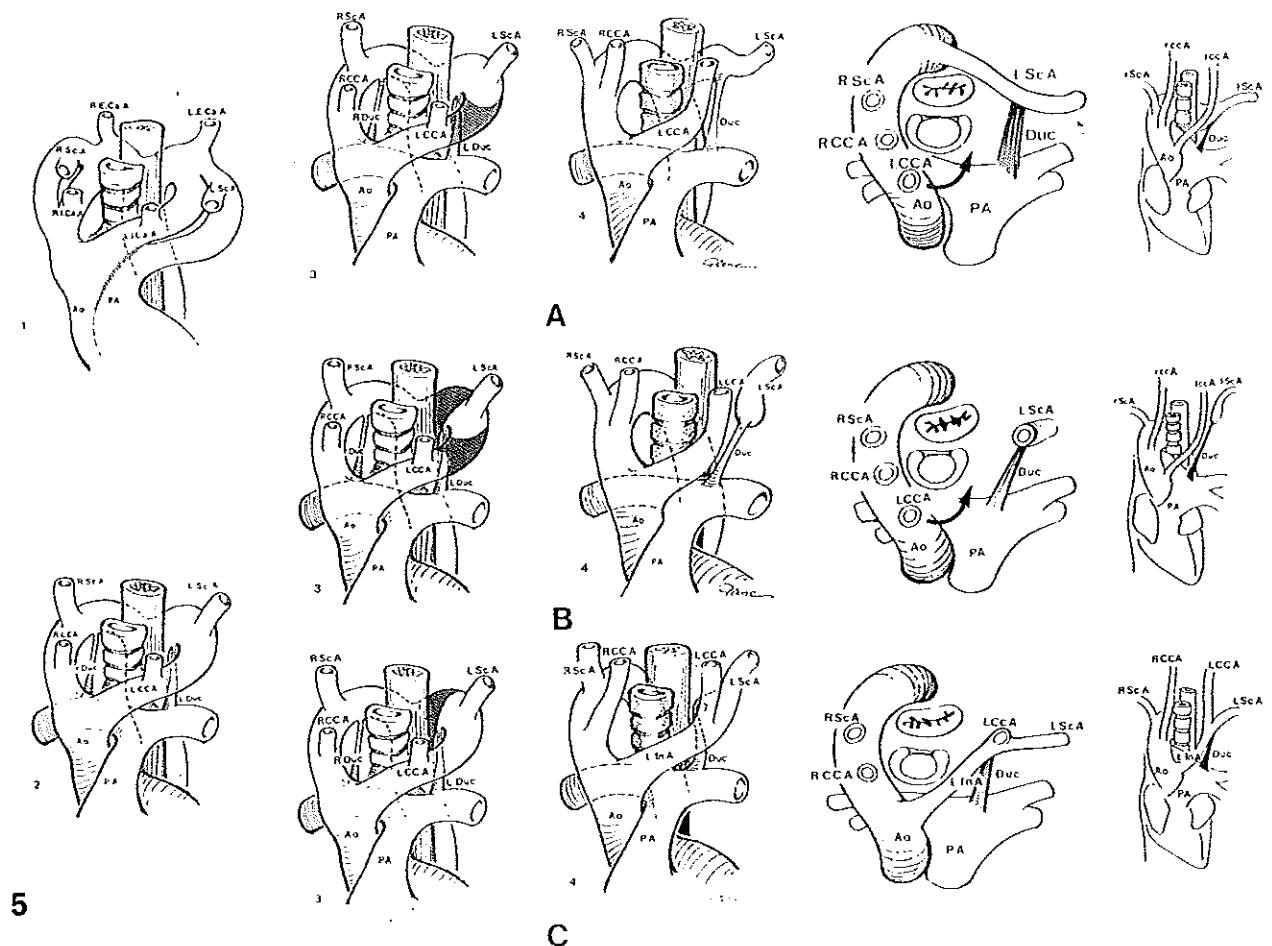


Fig. 5

Abnormal development of a right aortic arch. A) Regression occurs between left common carotid a. (LCCA) and left subclavian a. (LScA) with resultant aberrant subclavian a. B) Regression before and after left subclavian a. (LScA) with isolation of the subclavian a. C) Regression after the left subclavian a. (LScA) with formation of the mirror-image. Ao aorta PA pulmonary a. LICA left internal carotid a. RICA right internal carotid a. LECA left external carotid a. RECa right external carotid a. LCCA = left common carotid a. RCCA right common carotid a. LScA left subclavian a. RScA right subclavian a. Duct ductus

Développement anormal d'une crosse aortique droite. A) La régression a lieu entre l'a. carotide commune gauche (LCCA) et l'a. sub-clavière gauche (LScA) donnant une a. sub-clavière gauche aberrante. B) La régression a lieu avant et après l'a. sub-clavière gauche (LScA) avec exclusion de l'a. sub-clavière. C) La régression a lieu après l'a. sub-clavière gauche (LScA) avec formation d'une disposition en miroir de la crosse aortique

ching of the arch vessels [1, 2]. Interruption of the left arch occurs between the descending aorta and the left subclavian a. (Fig. 5b). A left innominate artery arises as the first branch followed by a right common carotid a. and a right subclavian a. The descending aorta is generally right sided, and the right arch with mirror-image usually has no upper retrooesophageal component.

Sometimes, the mirror-image arch crosses the midline towards the

left side high in the chest, with the presence of a localized posterior indentation of the oesophagus. Usually the interruption of the left arch occurs beyond the ligamentum arteriosum that is left sided, with no subsequent vascular ring. However in rare cases, regression of the left aortic arch may occur between the left subclavian and the ligamentum arteriosum which connects the left pulmonary a. to the upper descending aorta, producing a vascular ring.

In the third type (Fig. 5c), which

is the less common, the left aortic arch is found proximally between the left common carotid and the left subclavian aa., and distally behind the attachment of the left ductus [11]. The left subclavian a. has no connection with the aorta but is attached to the left pulmonary trunk by a left ligamentum arteriosum. This situation may lead to a subclavian steal syndrome.

Many intracardiac anomalies have been described associated with a right aortic arch [6]: tetralogy of

Fallot, ventricular septal defect, pulmonary stenosis, common atrioventricular canal, complete transposition. They are present in 80% of patients with the mirror-image type (type II). Most patients with an aberrant subclavian a. (type I) have no heart disease. When no posterior defect is visualized on the oesophagogram, the probability of associated cardiac disease is high.

Some chest x-ray findings suggest right aortic arch: widened superior mediastinal silhouette, absence of the aortic knob on the left side, deviation of the trachea to the right with an indentation on its right border and the presence of the descending aorta on the right of the spine [1, 8, 9]. However, in infants, the volume of the thymus gland complicates the localization of the aortic knob. Moreover, when a large diverticulum extends laterally beyond the left border of the mediastinum, it may take the appearance of a normal left-sided aortic knob.

When multiple investigations are used, aortography favours precise delineation of the right arch and of the branching pattern of the

supraaortic vessels. In our 2 cases, preoperative coronary angiography failed to diagnose the right aortic arch because the opacification of the aorta was limited to the supravalvular portion. Thus, the left common carotid a. was not recognized as the first branch arising from the aorta.

Surgical correction of right aortic arch is indicated only for those anomalies that are symptomatic [8, 9]. The operation consists in division of the constricting vascular ring or displacement of the aberrant vessels, which allows mobilization of the oesophagus from its mediastinal bed. Reconstruction of the divided left subclavian a. prevents the development of a subclavian steal phenomenon.

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