Chapter 3.14 Aortic arch interruption

Definition

The aortic arch is described as three segments: proximal, distal and isthmus. The proximal component extends from the take-off of the innominate artery to the left common carotid artery. The distal component extends from the left common carotid artery to take-off of the left subclavian artery. The segment of the aorta connecting the distal aortic arch to the juxtaductal region of the descending aorta is termed the isthmus. This complex composite of segments introduces a risk of developmental anomalies in the form of interruptions at the various junction points. Aortic arch interruption is characterized by complete lack of anatomic continuity between the aortic arch or isthmus and the descending thoracic aorta. In aortic arch atresia, with identical pathophysiology and hemodynamics, there is anatomic continuity between the two segments, represented by an imperforate fibrous strand of various lengths. Three anatomic types of aortic arch interruption have been described:

- *Type A*: the interruption is distal to the left subclavian artery.
- *Type B*: the interruption is between the left common carotid artery and the left subclavian artery.
- *Type C*: the interruption is between the innominate artery and the left common carotid artery.

Associated anomalies

Patent ductus arteriosus, ventricular septal defect, actual or potential systemic left ventricular outflow tract obstruction, bicuspid aortic valve, double outlet right ventricle, univentricular heart with discordant ventriculoarterial connection, aortopulmonary window, truncus arteriosus, atrial isomerism. Patent ductus arteriosus is always present. The ventricular septal defect is nearly always present, in the majority of cases of malalignment-type, subpulmonic, because of posterior malalignment of the ventricular septum. Subaortic stenosis is also generally present, due to the posterior malalignment of the ventricular septum.

Surgical options

Two surgical approaches are available:

- *two stages*, with repair of the interrupted aortic arch and pulmonary artery banding, followed by later closure of the ventricular septal defect;
- *single stage*, with simultaneous repair of the interrupted aortic arch and a patch closure of the ventricular septal defect.

In the absence of associated lesions other than patent ductus arteriosus and ventricular septal defect, primary repair by direct anastomosis of the arch with closure of the ventricular septal defect is the preferred surgical approach. The main reason is that pulmonary artery banding might promote or aggravate subaortic stenosis in patients with a malalignment ventricular septal defect. Although the primary repair is physiologically corrective, it should not be viewed as fully curative due to the high incidence of significant late obstruction of the left ventricular outflow tract.

Left ventricular outflow tract obstruction: The morphology of left ventricular outflow tract obstruction with interrupted aortic arch varies, and therefore, surgical management also varies according to the specific circumstances.

Pre-operative information

CT and MRI investigations (Figs. 3.14.1 and 3.14.2) allow the precise identification of aortic arch interruption type, distance between proximal and distal segments, size of the patent ductus arteriosus, narrowest dimension of the left ventricular outflow tract, and association of other malformations potentially interfering with the surgical planning, such as intracardiac defects, the presence of an anomalous retro-esophageal right subclavian artery, the presence of a right aortic arch.

The cardiac MRI protocol used for the evaluation of infants with aortic arch interruption is the following:

- 3-plane localizing images,
- 2-dimensional axial Time-of-flight Angiography (see appendix in the "Introduction"),
- ECG-gated cine steady-state free precession sequences in 2-chamber, 4-chamber planes, and ventricular short axis for the quantitative assessment of ventricular dimensions, function, mass and stroke volume, as illustrated in the appendix of the "Introduction",
- ECG-gated cine steady-state free precession sequence to visualize the left ventricular outflow tract and the aortic valve,
- gadolinium-enhanced 3-dimensional MRI for the evaluation of the aortic arch, brachiocephalic arteries and patent ductus arteriosus.

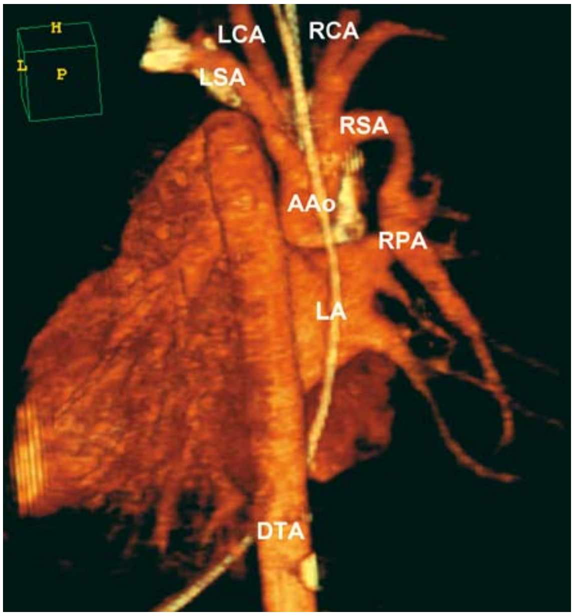

Fig. 3.14.1. Aortic arch interruption. CT scan posterior coronal view, volume rendering imaging, showing type A aortic arch interruption, distal to the left subclavian artery, in a neonate with anomalous origin of the right pulmonary artery from the right subclavian artery (*AAo* ascending aorta, *DTA* descending thoracic aorta, *LA* left atrium, *LCA* left carotid artery, *LSA* left subclavian artery, *RCA* right carotid artery, *RPA* right pulmonary artery, *RSA* right subclavian artery) (photograph courtesy of Dr. Mohammed Tawil)

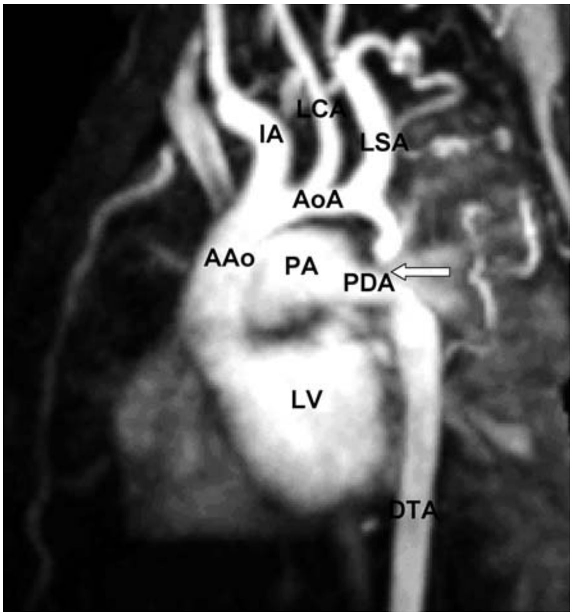

Fig. 3.14.2. Aortic arch interruption. MR angiography showing the aortic arch interruption (arrow), with the distal systemic circulation perfused through a patent ductus arteriosus (*AAo* ascending aorta, *AoA* aortic arch, *DTA* descending thoracic aorta, *IA* innominate artery, *LCA* left carotid artery, *LSA* left subclavian artery, *LV* left ventricle, *PA* pulmonary artery, *PDA* patent ductus arteriosus)

Fig. 3.14.3. Aortic arch interruption: status post repair. CT scan in the same patient of Fig. 3.14.1 with left oblique posterior sagittal view, volume rendering imaging, showing the end-to-side anastomosis (arrow) used to reconstruct the aortic arch continuity one year after surgery (*AAo* ascending aorta, *DTA* descending thoracic aorta) (photograph courtesy of Dr. Mohammed Tawil)

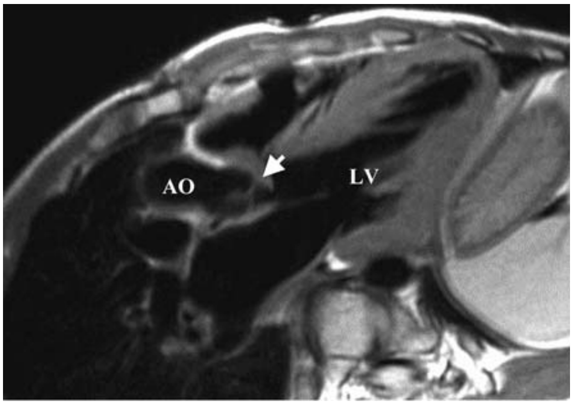

Fig. 3.14.4. Aortic arch interruption with ventricular septal defect, status post repair. MRI fast spin echo 3-chamber view parallel to the left ventricular outflow tract. Note the severe subaortic stenosis due to posterior infundibular deviation (arrow) (*Ao* aorta, *LV* left ventricle)

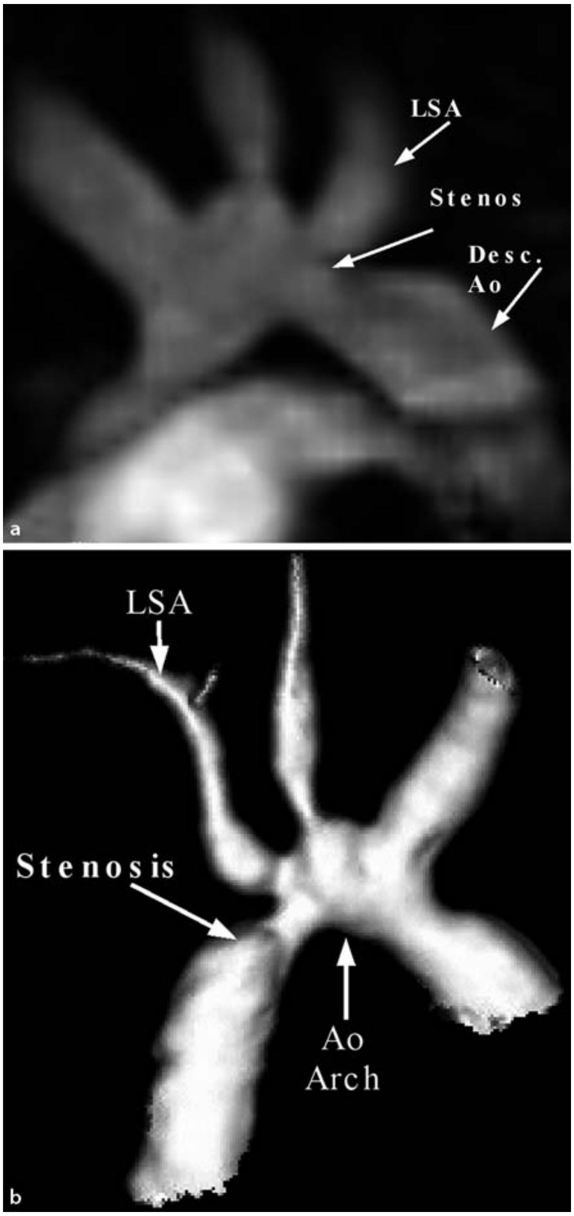

Fig. 3.14.5. Aortic arch interruption, status post surgery with direct anastomosis. MR angiography maximal intensity projection reconstruction (**a**) and volume rendering reconstruction (**b**) showing normal brachiocephalic arteries and significant aortic isthmus narrowing (*Ao Arch* aortic arch, *Desc Ao* descending thoracic aorta, *LSA* left subclavian artery)

▮ Potential complications

Residual pressure gradient at the level of the aortic arch anastomosis and/or at the left ventricular outflow tract, residual ventricular septal defect, complete atrioventricular block, left bronchial obstruction.

Left bronchial obstruction: The left main bronchus passes under the aortic arch. If a direct anastomosis is performed without adequate mobilization of the ascending and descending aorta, a bowstring effect over the left main bronchus may result. This is manifested by air trapping in the left lung with hyperexpansion, as seen on chest radiography and confirmed by bronchoscopy. Surgical management may require an ascending-to-descending aortic conduit after division of the arch.

▮ Post-operative follow-up

The role of CT scan (Fig. 3.14.3) and MRI after surgery for aortic arch interruption is to evaluate residual or recurrent anatomic and hemodynamic problems, such as aortic arch obstruction or aneurysm formation. However, other abnormalities (e.g., left ventricular outflow tract obstruction, aortic valve stenosis or regurgitation, residual ventricular septal defect, left ventricular size and function) should be examined as well. CT scan and MRI are also useful in case of suspected post-operative left bronchial compression.

The following protocol is suggested in the post-operative evaluation of patients with arch interruption:

- 3-plane localizing images,
- 2-dimensional axial Time-of-flight Angiography (see appendix in the "Introduction"),
- ECG-gated cine steady-state free precession and fast spin echo para-sagittal,

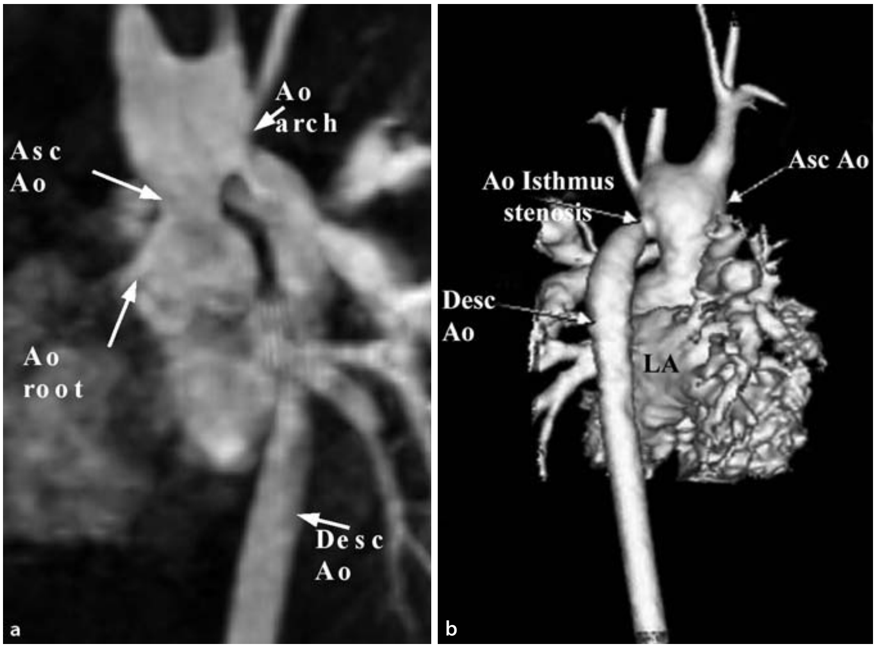

Fig. 3.14.6. Truncus arteriosus and aortic arch interruption, status post repair. MR angiography MIP reconstruction (**a**) and volume rendering reconstruction (**b**) showing an aortic arch aneurysm, mild ascending aorta stenosis and significant aortic isthmus obstruction (*Asc Ao* ascending aorta, *Ao arch* aortic arch, *Ao root* aortic root, *Desc Ao* descending thoracic aorta, *LA* left atrium)

coronal and additional oblique plane (if needed) to visualize the aortic arch,

- ECG-gated cine steady-state free precession sequences in 2-chamber, 4-chamber planes, and ventricular short axis for the quantitative assessment of ventricular dimensions, function, mass and stroke volume as illustrated in the appendix of the "Introduction",
- ECG-gated cine steady-state free precession and fast spin echo sequence parallel to the left ventricular outflow tract (Fig. 3.14.4),
- ECG-gated phase velocity contrast (PVC-MRI) sequences perpendicular to the ascending aorta, aortic arch, isthmus, proximal and distal descending aorta. Additional flow measurements, based on clinical relevance, such as assessment of potential aortic valve regurgitation or to quantitate the intracardiac shunt in case of suspected residual ventricular septal defect, are sometime required as well,
- gadolinium-enhanced 3-dimensional MRI for the evaluation of the aortic arch and brachiocephalic arteries (Figs. 3.14.5 and 3.14.6).

References

Akdemir R, Ozhan H, Erbilen E, Yazici M, Gunduz H, Uyan C (2004) Isolated interrupted aortic arch: a case report and review of the literature. Int J Cardiovasc Imaging 20:389–392

Cinar A, Haliloglu M, Karagoz T, Karcaaltincaba M, Celiker A, Tekinalp G (2004) Interrupted aortic arch in a neonate: multidetector CT diagnosis. Pediatr Radiol 34:901–903

Corno AF, Lam J, Becker AE, Marcelletti C (1981) Aortic arch interruption. G Ital Cardiol 11:679–685

Davotoglu V, Soydinc S, Sirikci A, Dinckal H, Akdemir I (2004) Interrupted aortic arch in an adolescent male. Can J Cardiol 20:1367–1368

Dorfman AL, Geva T (2006) Magnetic resonance imaging evaluation of congenital heart disease: conotruncal anomalies. J Cardiovasc Magn Reson 8:645–659

Goo HW, Park IS, Ko JK, Kim YH, Seo DM, Yun TJ, Park JJ, Yoon CH (2003) CT of congenital heart disease: normal anatomy and typical pathologic conditions. Radiographics 23:S147–S165

Johnson TR, Goldmuntz E, McDonald-McGinn DM, Zackai EH, Fogel MA (2005) Cardiac magnetic resonance imaging for accurate diagnosis of aortic arch anomalies in patients with 22q11.2 deletion. Am J Cardiol 96:1726–1730

Maier JM, Scheffold N, Cyran J (2005) Primary diagnosis of an interrupted aortic arch in a 65-year old woman with hypertension. Dtsch Med Wochenschr 130:2893–2895

Morriss J, Moreland J, Burkhart H, Kao S (2007) Presurgical evaluation of interrupted aortic arch with 3-dimensional reconstruction of CT images. Ann Thorac Surg 84:299

Taylor AM, Jones R, Derrick G (2006) Post-mortem magnetic resonance imaging provides the diagnosis following aortic rupture in an infant with treated aortic interruption. Cardiol Young 16:81–83

Vriend JW, Lam J, Mulder BJ (2004) Complete aortic arch obstruction: interruption or aortic coarctation? Int J Cardiovasc Imaging 20:393–396

Yoo SJ, Choi HY, Park IS, Hong MG, Kim SH (1991) Distal aortopulmonary window with aortic origin of the right pulmonary artery and interruption of the aortic arch (Berry syndrome): diagnosis by MR imaging. Am J Roentgenol 157:835–836

Wong MN, Chang LG, Sim KH (2007) Interrupted aortic arch and aortopulmonary window demonstrated on 64-slice multidetector computed tomography angiography. Heart 93:95

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