

# How Could Aortic Atresia With Interrupted Aortic Arch Survive? About a Neonatal Repair on Two Ventricles



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Aortic atresia with interrupted aortic arch (IAA) is an exceptional condition. In absence of associated lesion, the flow to the brain and the retrograde flow to the coronary arteries can only come from vessels on the descending aorta. This flow needs to be large enough to supply the brain and the myocardium. The only IAA type compatible with survival is type C where the flow from descending aorta could be insured by the left carotid and the left vertebral artery, branch of the left subclavian artery. Only one such a case was described in the literature. All the other surviving cases had an associated lesion including: a double aortic arch, an aorto-pulmonary window (AP window), an aberrant right subclavian artery or a bilateral patent ductus arteriosus (PDA). We report a case of aortic atresia with IAA type B associated with a double aortic arch that underwent a successful bi-ventricular one-stage neonatal Norwood-Rastelli repair and is doing well after 18 months.

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## BACKGROUND

As shown in Fig. 1, in absence of associated lesion, the only way for patients with aortic atresia and interrupted aortic arch (AA-IAA) to survive is through vessels coming from the descending aorta (left vertebral artery and/or left carotid artery) flowing to the circle of Willis to perfuse the brain and then the hypoplastic aorta and the coronary arteries through retrograde flow via the right carotid artery and/or a right vertebral artery. Therefore AA-IAA type A can never survive. Equally, AA-IAA type B cannot survive, as the only flow to the Willis circle is the left vertebral artery, which will be insufficient to perfuse together the brain and the coronary arteries. Only AA-IAA type C may survive because the flow to the circle of Willis is generated together by the left carotid and the left vertebral artery. Indeed, one such case of AA-IAA type C without associated lesion, has survived.<sup>1</sup>

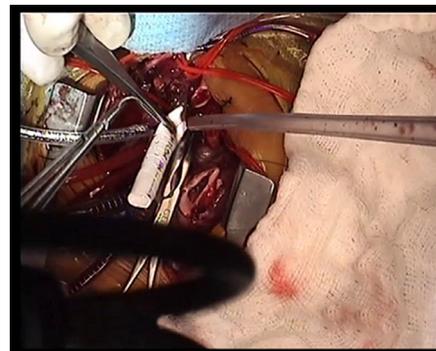
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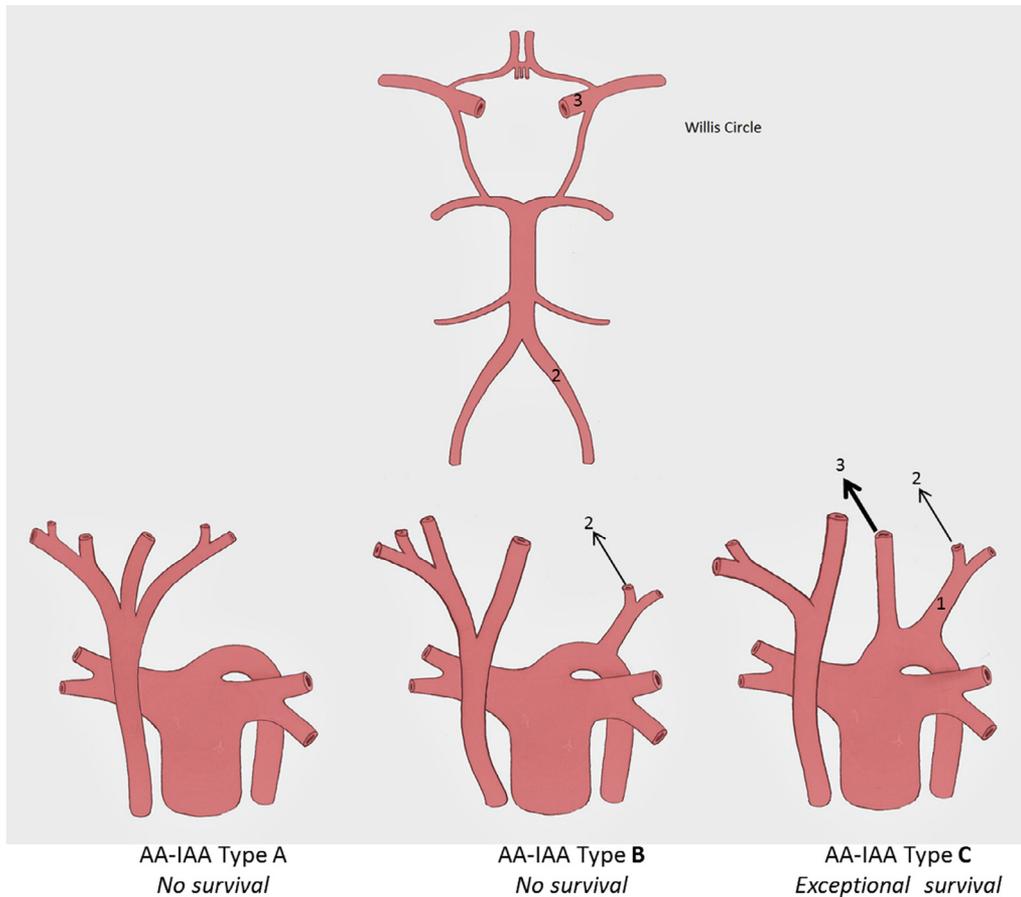
Neonatal Norwood-Rastelli repair (Yasui-like) for AA-IAA type B & double aortic arch.

## Central Message

Aortic atresia (AA) and interrupted aortic arch (IAA) without associated lesion can exceptionally survive. A neonatal biventricular repair of AA-IAA type B, with double aortic arch, is reported.

## CASE REPORT

A 5-day female, 2.8 kg, under prostaglandin, was diagnosed at echocardiography with: complete aortic valve atresia (AA), IAA type B, 2 mm ascending aorta, large patent ductus arteriosus (PDA), left subclavian artery (LSCA) from descending aorta, large cono-truncal ventricular septal defect (VSD) and two good-size ventricles. CT angiography revealed a double aortic arch (Fig. 2) formed by a 7 mm large posterior vessel arising from the descending aorta and crossing posterior to the tracheo-esophageal tract to end in the right SCA. At 11 days of age, a Norwood-Rastelli biventricular repair (Yasui-like) was performed (see Video). A small thymus was present. The hypothermic CPB was established with double cannulation: of the PDA and directly the left carotid for antegrade cerebral perfusion. The posterior aortic arch vessel was found on the descending aorta beyond the origin of the LSCA. It was divided, then sutured on the aortic side and doubly clipped on the other side allowing it to retract behind the tracheo-esophageal tract. The pulmonary trunk and the



**Figure 1.** AA-IAA types without associated lesion.

AA, aortic atresia; IAA, interrupted aortic arch; 1, left subclavian artery; 2, left vertebral artery; 3, left carotid artery.

hypoplastic ascending aorta were transected and anastomosed side-by-side. A 6 mm Goretex [W.L. Gore & Associate Inc., Flagstaff, AZ] conduit was interposed between the transverse arch and the descending aorta. The double aorto-pulmonary root was then anastomosed to the inferior side of the Goretex conduit. The 8 mm large venetricular septal defect (VSD) was re-routed to the pulmonary artery, using a bovine pericardium patch. A 12 mm Contegra conduit [Medtronic Inc, Minneapolis, MN] was implanted between the pulmonary artery bifurcation and the right infundibulotomy. Separation from CPB was easily obtained. The chest was left open and the sternum closed 2 days later. A postoperative CT angiogram showed compression of the distal trachea. A reoperation through right thoracotomy allowed removing of the entire remnant of the posterior vessel along with the two clips. The patient was successfully extubated at day 12 and was discharged home 1 month after the first surgery. Eighteen months after surgery, the child is growing well. A control angiography showed moderate stenosis on the

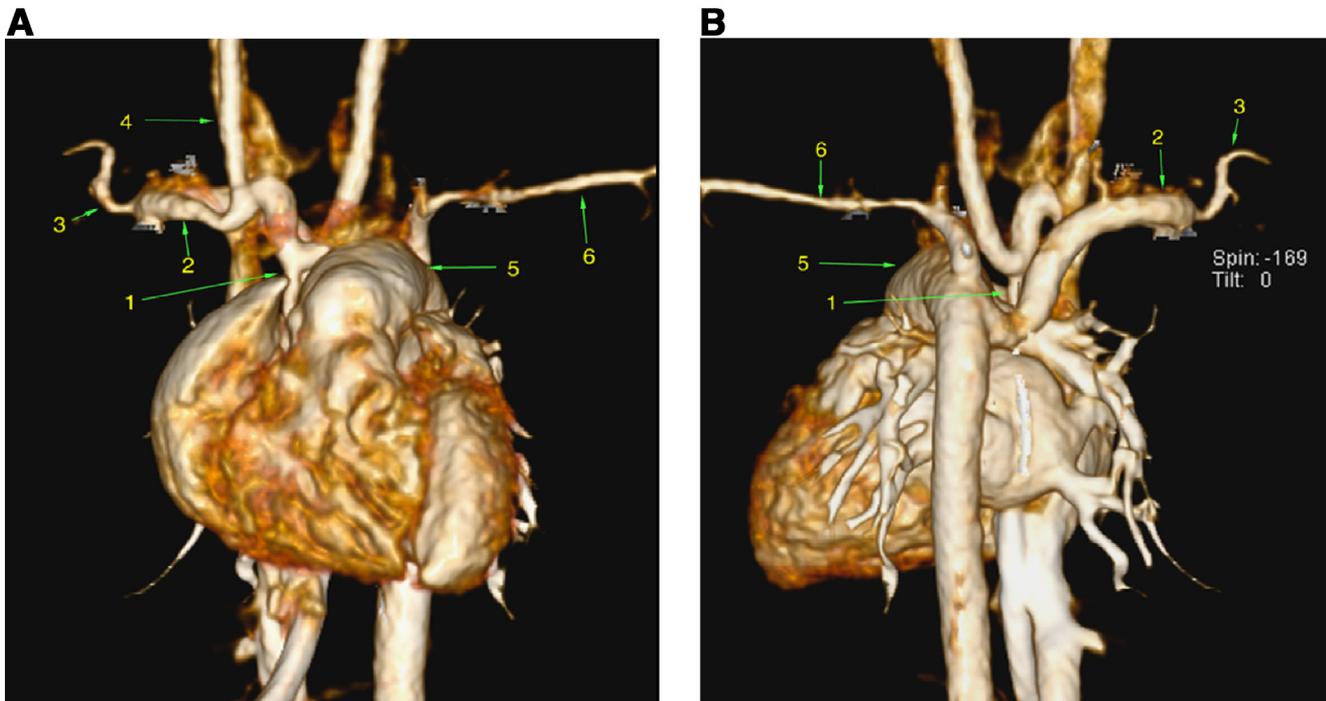
pulmonary bifurcation that was successfully dilated (see Video).

**COMMENTS**

The association of IAA and AA, with venetricular septal defect (VSD) and two viable ventricles is exceedingly rare. Norwood<sup>2</sup> successfully repaired for the first time an AA-IAA type C with bilateral PDA. Only one case without associated lesion (Table 1) is reported<sup>1</sup> in a patient with AA-IAA type C and a left brachio-cephalic artery arising from the descending aorta. The authors have not considered this aberrant left brachio-cephalic artery as an associated lesion and have considered this case as an equivalent of IAA type C.

All the other 12 cases published had an associated lesion (Table 1), in the form of a vessel providing a supplemental flow to the Willis circle or to the ascending aorta. The flow is exclusively “Willis circle dependent” with reverse flow to

## CONGENITAL – AORTIC ATRESIA WITH INTERRUPTED AORTIC ARCH



**Figure 2.** (A) CT angiogram, anterior view. (B) CT angiogram, posterior view.

1, aortic root, 2, posterior aortic arch ending onto the right subclavian artery; 3, right subclavian artery; 4, right internal carotid artery; 5, patent ductus arteriosus; 6, left subclavian artery.

the coronary arteries in 3 cases of aberrant right SCA.<sup>3–5</sup> The other 9 cases have a coronary flow not dependent on the Willis circle: bilateral PDA in 2 cases,<sup>2,6</sup> aorto-pulmonary window (AP window) in 4<sup>7–10</sup> and double aortic arch in 3.<sup>11,12</sup> To notice that we have found only one case of

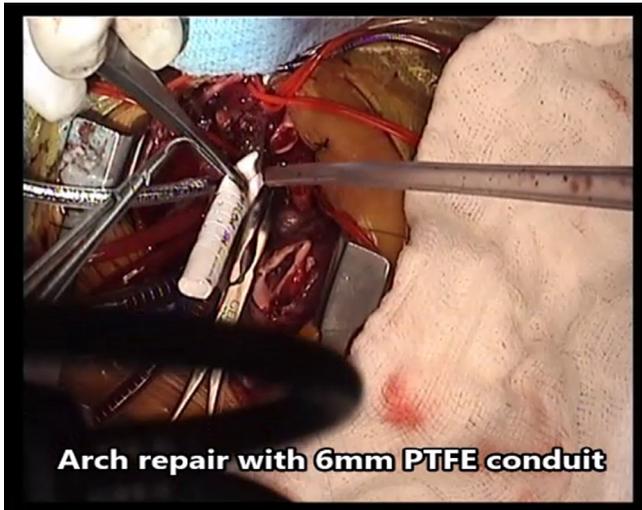
hypoplastic left heart syndrome—AA with IAA who survived a Norwood operation.<sup>3</sup> The one-stage neonatal biventricular Norwood-Rastelli repair is the optimal surgical technique. This can be safely achieved today following advances in neonatal cardiac surgery.

**Table 1.** Thirteen Cases of AA-IAA. Only Case 1, AA-IAA Type C, Had No Real Associated Lesion. Four Cases Had a Willis Circle Dependent Circulation. Nine Cases Had an Obvious Associated Lesion

| AA-IAA Type | Associated Lesion  | Willis Circle Dependent Circulation | Survival | Authors                              |
|-------------|--------------------|-------------------------------------|----------|--------------------------------------|
| C           | None               | Yes                                 | Yes      | Malankar et al 2017 <sup>1</sup>     |
| B           | Aberrant right SCA | Yes                                 | Yes      | Tannous et al 2006 <sup>5</sup>      |
| B           | Aberrant right SCA | Yes                                 | Yes      | Careddu et al 2015 <sup>4</sup>      |
| B           | Aberrant right SCA | Yes                                 | Yes      | Dibardino et al 2005 <sup>3</sup>    |
| C           | Bilateral PDA      | No                                  | Yes      | Norwood et al 1981 <sup>2</sup>      |
| C           | Bilateral PDA      | No                                  | No       | Decaluwe et al 2005 <sup>6</sup>     |
| B           | AP window          | No                                  | No       | Rosenquist et al 1974 <sup>7</sup>   |
| B           | AP window          | No                                  | No       | Redington et al 1991 <sup>8</sup>    |
| B           | AP window          | No                                  | No       | De Caro et al 1998 <sup>9</sup>      |
| B           | AP window          | No                                  | No       | Yew et al 2005 <sup>10</sup>         |
| B           | Double aortic arch | No                                  | ?        | Agnoletti et al 2001 <sup>11</sup>   |
| B           | Double aortic arch | No                                  | No       | Baker-Smith et al 2010 <sup>12</sup> |
| B           | Double aortic arch | No                                  | Yes      | Our case                             |

**SUPPLEMENTARY MATERIAL**

The following is the supplementary data to this article:



**Video 1.** Neonatal Norwood-Rastelli repair (Yasui-like) for AA-IAA type B & double aortic arch.

**REFERENCES**

1. Malankar DP, Glatz AC, Weinberg PM, et al: Biventricular repair in interrupted aortic arch type C with aortic atresia. *World J Pediatr Congenit Heart Surg* 2017;2150135117715211
2. Norwood WI, Stellan GJ: Aortic atresia with interrupted aortic arch: Reparative operation. *J Thorac Cardiovasc Surg* 81:239–244, 1981
3. Dibardino DJ, Heinle JS, Andropoulos DA, et al: Aortic atresia and type B interrupted aortic arch: Diagnosis by physiologic cerebral monitoring. *Ann Thorac Surg* 79:1758–1760, 2005
4. Tannous HJ, Moulick AN, Jonas RA: Interrupted aortic arch and aortic atresia with circle of Willis-dependent coronary perfusion. *Ann Thorac Surg* 82:11–13, 2006
5. Careddu L, Oppido G, Ialonardi M, et al: Aortic atresia with interrupted aortic arch: a combination incompatible with life? *Interact Cardiovasc Thorac Surg* 21:272–273, 2015
6. Decaluwe W1, Delhaas T, Gewillig M: Aortic atresia, interrupted aortic arch type C perfused by bilateral arterial duct. *Eur Heart J* 26:2333, 2005
7. Rosenquist GC, Taylor JF, Stark J: Aortopulmonary fenestration and aortic atresia. Report of an infant with ventricular septal defect, persistent ductus arteriosus, and interrupted aortic arch. *Br Heart J* 36:1146–1148, 1974
8. Redington AN1, ML Rigby, Ho SY, et al: Aortic atresia with aortopulmonary window and interruption of the aortic arch. *Pediatr Cardiol* 12:49–51, 1991
9. De Caro EI, Pongiglione G, Ribaldone D: Interruption of the aortic arch, ventricular septal defect, aortic atresia and aortopulmonary fistulous communication. *Int J Cardiol* 65:19–21, 1998
10. Yew G, Coleman D, Calder L: Aortic valvar atresia, interrupted aortic arch, and quadricuspid pulmonary valve: A rare combination. *Pediatr Cardiol* 26:455–459, 2005
11. Agnoletti G1, Borghi A, Annecchino F: A rare form of interrupted aortic arch. *Ital Heart J* 2(3):228–230, 2001
12. Baker-Smith CM1, Milazzo AS, Frush DP, et al: Double aortic arch with aortic atresia and left-sided type B interruption. *Congenit Heart Dis* 5:316–320, 2010