INTRODUCTION: Aortic arch anomalies are associated with various types of heart defects ranging from ventricular septal defect to left heart hypoplasia. Decision about single- or two-stage biventricular repair in children with good left ventricle depends on child’s weight, type of associated heart defect and local conditions during surgery.

THE AIM OF THIS STUDY was to present a series of four cases with complex heart defects associated with interrupted or hypoplastic aortic arch and aortic valve atresia or hypoplasia, in whom anatomical conditions did not allow for single-stage biventricular correction and the two-stage repair with initial Norwood-type operation was performed.

CASE REPORTS: Two patients have a hypoplastic aortic arch associated with aortic valve atresia and ventricular septal defect, one has interrupted aortic arch with ventricular septal defect and one has interrupted aortic arch with atrial and ventricular septal defects. All patients had initial Norwood-type operations with right ventricle to pulmonary artery shunts and various modifications depending on heart anatomy. One child required angioplasty of RV-PA shunt with stent implantation 10 months after initial operation, in one case a B-T shunt was performed 19 months after Norwood procedure. Biventricular correction was performed in all four cases an average of 18.25 months after Norwood procedure. In two of the cases a pulmonary homograft, in one contegra conduit and in one matrix valved graft was used for right ventricle outflow tract reconstruction. All children are in good condition with laminar flow through the neoaorta and pulmonary graft and after average 20(SD±6.5) months of follow up with no need of reinterventions and reoperations. One child required pharmacotherapy (Salbutamol) because of advance atrioventricular block (II/III degree).

CONCLUSIONS: Double-stage biventricular repair in cases of interrupted or hypoplastic aortic arch associated with aortic valve atresia or hypoplasia and ventricular septal defect, with initial Norwood-type operation ensures good postoperative effect and is an interesting alternative for single-stage correction in cases of significant left ventricle outflow tract obstruction and low body weight neonates with poor local condition unsuitable for arch reconstruction.