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Brief Report

Modified Nikaidoh procedure in a patient with transposition of the great arteries, ventricular septal defect, and left ventricular outflow tract obstruction with unusual coronary anatomy

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Abstract The Rastelli operation has been the most common procedure for the repair of transposition of the great arteries with ventricular septal defect and left ventricular outflow tract obstruction. A relatively recent approach is the Nikaidoh procedure. Despite the fact that it seems promising, the operation lacks long-term follow-up data. It has been postulated that patients with anomalous coronary arteries are high-risk candidates for the Nikaidoh procedure and its modifications. In this report, we present the case of a patient with transposition of the great arteries with remote restrictive ventricular septal defect and left ventricular outflow tract obstruction with coronary anomaly – with the right coronary artery originating from the left anterior descending coronary artery and crossing the right ventricular outflow tract – who underwent successful modified Nikaidoh operation.

Keywords: Paediatric cardiac surgery; Rastelli procedure; Réparation à l'Etage ventriculaire; Nikaidoh procedure

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A combination of transposition of the great arteries with ventricular septal defect and left ventricular outflow tract obstruction represents a minor group among congenital cardiac defects; [1–8] however, the combination is a challenge among paediatric cardiac surgeons. Different surgical techniques such as the Rastelli procedure, Réparation à l'Etage ventriculaire and the Metras modification, and Nikaidoh operation and its modifications were defined; however, the Rastelli procedure has been the most common surgical treatment up to date.[1–8] However, long-term results of the Rastelli procedure, Réparation à l'Etage ventriculaire, and the Metras modification carry risks of considerable mortality and morbidity, [1–8] and Nikaidoh operations and its modifications still require long-term follow-up results in larger series.

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Despite the limited number of patients and follow-up data, the originally described Nikaidoh procedure in 1984,[1] and its various modifications presented in recent years,[2–5] seem promising. The literature consists of reports indicating unusual coronary anatomy as a contraindication for the Nikaidoh operation.[3,4] In this report, we present the case of a patient with the transposition of the great arteries, remote ventricular septal defect, and left ventricular outflow tract obstruction with coronary anomaly, who underwent successful modified Nikaidoh procedure.

Case report

The patient was a 3-year-old male weighing 16 kilograms, diagnosed with transposition of the great arteries, ventricular septal defect, and left ventricular outflow tract obstruction. The ventricular septal defect was remote and restrictive. In his history, there was a modified Blalock–Taussig shunt that was performed through median sternotomy with a 4-millimetre

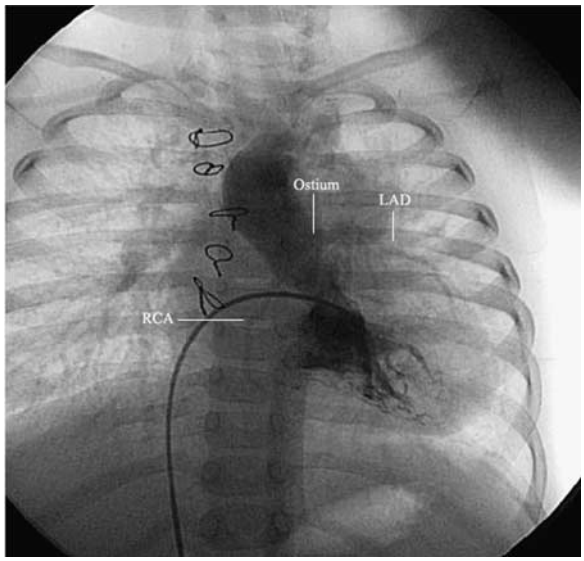

Figure 1.
The cardiac catheterisation showing the right coronary artery originating from the left anterior descending artery. RCA = right coronary artery; LAD = left anterior descending coronary artery.

expanded polytetrafluoroethylene graft when he was 3 months old. Despite the fact that echocardiography and angiography confirmed the functioning shunt, the room-air oxygen saturation was 65%. In addition, cardiac catheterisation indicated unusual coronary pattern: the right coronary artery originated from the left anterior descending artery (Fig 1) and normal origin circumflex coronary artery. Most probably due to post-stenotic dilatation, the main pulmonary artery and both branches were large and revealed a McGoon value above 2.

Surgical technique

The heart was accessed through median sternotomy. There were extensive adhesions, and care was taken to free the heart and vascular structures from these adhesions. Cardiopulmonary bypass was initiated with cannulation of the distal ascending aorta and superior and inferior caval veins. Cardioprotection was achieved with moderate hypothermia (28 degrees Celcius) and cold intermittent antegrade blood cardioplegia. The modified Blalock–Taussig shunt was ligated. The pulmonary arteries were large. Despite the fact that the coronary anatomy was identified before the surgery with cardiac catheterisation, the right coronary artery was accidentally injured during the aortic root preparation from the right ventricle (Fig 2) because of extensive adhesions that occurred after his first operation. The right coronary artery was anastomosed end to end with an 8.0 polyprolene suture. Repair of the right coronary artery was easy because of the large size of the artery in accordance with the child's age, as well as cyanotic status of the patient – that is, in

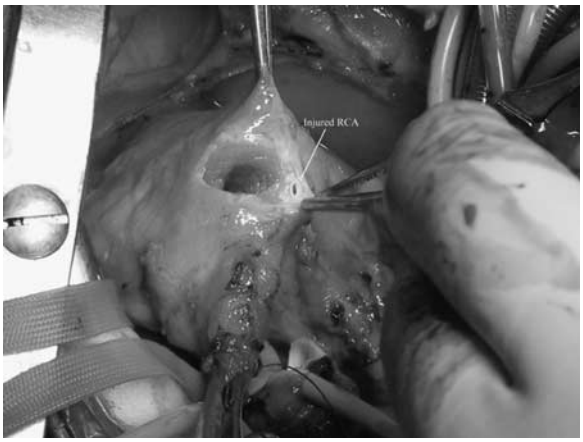

Figure 2.
The right coronary artery was accidentally injured during the aortic root preparation. RCA = right coronary artery.

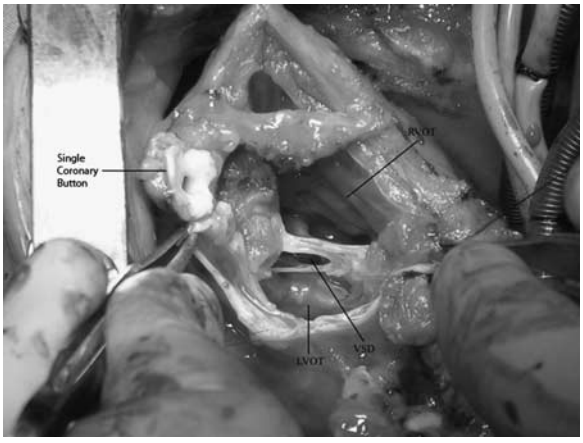

Figure 3.
Figure showing the pathology. RVOT = right ventricular outflow tract; LVOT = left ventricular outflow tract; VSD = ventricular septal defect.

cyanotic congenital cardiac disorders, the coronary arteries are enlarged. The circumflex coronary artery was not removed as a button, but was prepared free from the right ventricular wall to facilitate aortic translocation. The aortic root was completely dissected, leaving the left anterior descending coronary artery together with right coronary artery as a button. The pulmonary artery was dissected. The left ventricular outflow tract was hypoplastic with an annulus size of 4–5 millimetres (Fig 3). A "V"-shaped dacron patch was used to reconstruct the new outflow tract and close the ventricular septal defect (Fig 4). The left ventricular outflow tract was enlarged by incising the infundibular septum and trimming the hypertrophied muscle bands at the left ventricular outflow tract. Lecompte manoeuvre was performed. The aortic root containing the circumflex coronary artery was translocated with posterior rotation to the remodelled left ventricular

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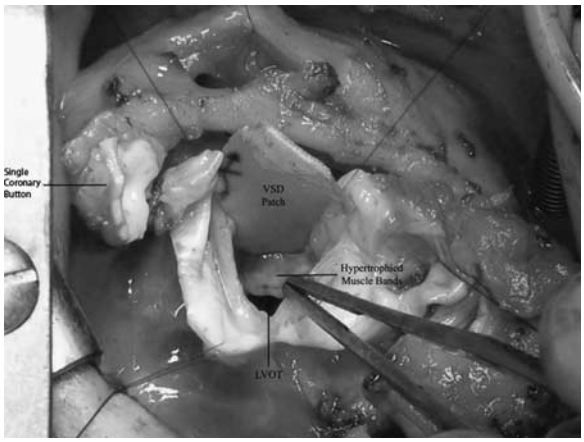

Figure 4.
A "V"-shaped dacron patch was used to reconstruct the new outflow tract and close the ventricular septal defect. LVOT = left ventricular outflow tract; VSD = ventricular septal defect.

outflow tract. The detached left anterior descending coronary artery was then reimplanted to the most suitable location on the translocated aorta. Despite the fact that pre-operative calculations indicated a conduit size of 16 millimetres for the reconstruction of the right venrtricular outflow tract, a Contegra valved conduit with a size of 14 millimetres was used to provide pulmonary flow due to the close anatomy of the right coronary artery crossing the right ventricular outflow tract to prevent right coronary artery distention and myocardial ischaemia.

The cardiopulmonary bypass and aortic cross-clamp times were 185 and 142 minutes, respectively. The patient experienced Wenkebach Type 2-like rhythm disturbances, and therefore we instituted ventricular pacing perioperatively. He was weaned off cardiopulmonary bypass with moderate dose inotropic support – 5 micrograms per kilogram per minute Dopamine, and 0.05 microgram per kilogram per minute Adrenalin. We did not come across coronary artery-related ischaemia symptoms, which might have resulted from the right coronary artery, thus confirming the successful repair of the injured right coronary artery. At post-operative echocardiography, the right ventricular outflow tract indicated a maximum of 20-millimetre mercury gradient through the Contegra and minimal aortic valve insufficiency. The patient was transferred to the intensive care unit and was extubated on the fourth post-operative day. The rhythm problem ceased spontaneously on the third day. The post-operative course was uneventful and he was discharged on day 10 without any problems. He was at New York Heart Association Functional Class I, and was symptom free in the follow-up period, with 100% oxygen saturation at room air for more than 8 months, without signs and symptoms of coronary ischaemia.

Comment

Transposition of the great arteries, ventricular septal defect, and left ventricular outflow tract obstruction pathology has long been treated with different techniques. Among them, since its first introduction in 1969, the Rastelli procedure is indisputably the most common method with long-term follow-up results.[2–9] The procedure has evolved over time with contributions from different authors for better outcomes.[10] However, long-term follow-up results of the Rastelli procedure have shown the unfeasibility of the method due to considerable rates of reoperations, rhythm disturbances, and low 20-year survival rate.[4–9]

In 1982, as an alternative to the Rastelli procedure, a new technique, the Réparation à l'Etage ventriculaire procedure, was introduced by Lecompte et al[11] In this method, the right ventricular outflow tract is reconstructed by directly anastomosing the pulmonary artery to the right ventricular outflow tract, and hence left valveless and without using a prosthetic conduit.[10] Metras repair is a modification of Réparation à l'Etage ventriculaire in which the pulmonary artery is still left valveless and connected to the right ventricle, but with an elongation segment obtained from the autologous aorta.[12] Valveless right ventricular outflow tract reconstructions may provide longer reoperation-free interval; however, inevitably, these patients will require a reintervention for the valveless pulmonary artery and right ventricular functions in the long run.

In 1984, Nikaidoh announced the latest technique for the treatment of this particular group of patients.[1] The method is a combination of the Ross, Konno, and Jatene procedures.[4] It provides anatomical alignment of the aorta over the left ventricle and better haemodynamic and physiologic performance at the left and right ventricular outflow tracts. The native aortic valve is transferred to the systemic circulation with anatomical alignment, and therefore it is believed that the longevity of the procedure will be better.[5]

Modifications of Nikaidoh differ from the original technique mainly on the basis of the type of the coronary transfers. In the original technique, aortic root is transferred en bloc to the left ventricular outflow tract, whereas in the modified techniques either left or right coronary artery or both are detached and reimplanted following aortic translocation to prevent coronary insufficiency.[2–4,13]

It is well known that the Rastelli, Réparation à l'Etage ventriculaire, or Metras operations do not require coronary transfer. However, during the Nikaidoh-type repairs, the coronary arteries are transferred either en bloc or separately. It has been accepted as a risk factor for increased mortality and morbidity in the conventional arterial switch operations, and thus some authors propose unusual

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coronary anatomy as a contraindication for the Nikaidoh procedure.[3,4] The reasons are mainly due to the high risk and aggressive nature of the Nikaidoh and its modifications. In the literature, only Hu et al[5] presented cases of two patients with coronary anomalies who underwent Nikaidoh procedures; however, the authors did not explain their techniques in these two particular cases in detail. As can be seen in the history of the Jatene procedures, increasing experience will eventually cease the high-risk nature of challenging coronary anatomy during the Nikaidoh operations.

The Nikaidoh procedure may include some limitations, and our case clearly shows why some authors considered coronary anomalies as relative contraindications to the Nikaidoh.[3,4] The coronary pattern should be extensively evaluated in the pre-operative period, as well as during the surgery. Our patient was 3 years old; however, if the patient was much smaller or even a neonate, repair of the injured coronary artery would not have been as easy or successful, and the procedure may have ended up with catastrophic results. However, Nikaidoh-type repairs are very rarely indicated in neonates or small children. In children with challenging coronary anatomies, such as intramural coronary course, one must not hesitate to proceed with alternative techniques such as Rastelli or Réparation à l'Etage ventriculaire procedures even though the Nikaidoh procedure was planned in the pre-operative period.

In the presented case, although a conduit size of 16 millimetres was required, a 14-millimetre-sized Contegra valved conduit was used for the right ventricular outflow tract reconstruction because of the close anatomy of the right coronary artery to the right ventricular outflow tract. This did not lead to low oxygen saturation because the child was not very small. It also provided distension-free right coronary artery and prevented myocardial ischaemia. Thus, the child has been at New York Heart Association Functional Class I, without signs and symptoms of coronary ischaemia, with 100% oxygen saturation at room air.

Conclusion

The modified Nikaidoh procedure is one of the surgical options for transposition of the great arteries, ventricular septal defect, and left ventricular outflow tract obstruction, providing physiologic cardiac haemodynamics. It has low reoperation rate for the right and left ventricular outflow tracts in the mid-term follow-up. Its long-term benefits need to be evaluated with larger number of patients and longer follow-up data. We believe that the procedure can be performed safely in children with this particular congenital cardiac anatomy, even in the presence of a coronary anomaly in selected patients. However, the coronary anatomy must be explicitly studied before surgery, and coronary arteries must be very carefully prepared during surgery. This single case experience shows the feasibility of the procedure with increasing experience.

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