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Original Article

Intensive care management of neonates with d-transposition of the great arteries and common arterial trunk

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Abstract Although mortality rates for patients with d-transposition of the great arteries remain quite low, these patients have a unique circulation that requires careful management in the peri-operative period. Despite the improved mortality for patients with common arterial trunk, the course in the intensive care unit is remarkable for significant morbidity and utilisation of significant resources. Pre-operative patient management focuses on balancing competing circulations, pulmonary and systemic, which exist in parallel rather than in series, as in the normal circulation. Post-operative patient management in both lesions focuses on optimising systemic output, respiratory status, and mitigating the effects of cardiopulmonary bypass. In this article, we review pre- and post-operative intensive care management in neonates with d-transposition of the great arteries and common arterial trunk.

Keywords: D-transposition of the great arteries; truncus arteriosus; peri-operative care; intensive care; 22q11 deletion

D-transposition of the great arteries

D-TRANSPOSITION OF THE GREAT ARTERIES IS THE most common form of cyanotic congenital heart disease, accounting for 5–9% of all cardiac malformations.[1,2] It is defined by the presence of ventriculoarterial discordance with the aorta arising anteriorly from the morphologic right ventricle and the pulmonary artery arising posteriorly from the morphologic left ventricle. This arterial configuration results in parallel systemic and pulmonary circulations that, without intervention, are often fatal, with a mortality rate of 90% within the first year of life.[3,4] Current pre-operative and operative advances have favourably changed patient outcomes, with long-term survival approaching 97%.[4–7]

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Physiologic and anatomic considerations

The discordant relationship between the ventricles and great arteries in d-transposition of the great arteries creates parallel systemic and pulmonary circulations. This results in significant systemic hypoxaemia due to ineffective delivery of blood to the pulmonary circulation for oxygenation and increased cardiac workload.

The degree of hypoxaemia in d-transposition of the great arteries is dependent on the presence and patency of intercirculatory communications. These communications may be intracardiac – patent oval foramen, atrial septal defect, or ventricular septal defect – or extracardiac – patent arterial duct or bronchopulmonary collaterals. Effective systemic and pulmonary blood flow is dependent on the presence of the aforementioned communications and should be considered in the context of the following principles:[8]

- Anatomic left-to-right shunting (effective systemic blood flow): net blood flow entering the systemic circulation from the pulmonary circulation;

- Anatomic right-to-left shunting (effective pulmonary blood flow): net blood flow entering the pulmonary circulation from the systemic circulation;
- Physiologic left-to-right shunting: volume of recirculating pulmonary venous blood flow;
- Physiologic right-to-left shunting: volume of recirculating systemic venous blood flow.

The amount of intercirculatory mixing is the volume of anatomic left-to-right shunting (effective systemic blood flow), which is equal to the volume of anatomic right-to-left shunting (effective pulmonary blood flow).

Several anatomic considerations should be borne in mind when evaluating infants with d-transposition of the great arteries with respect to intercirculatory mixing and effective flow of blood. Infants with d-transposition of the great arteries and a ventricular septal defect may present with mild cyanosis due to increased intercirculatory mixing and may also present with signs of increased pulmonary blood flow and congestive cardiac failure. However, the presence of a ventricular septal defect does not ensure adequate intercirculatory mixing and these patients may still require pre-operative interventions to establish effective mixing.

Conversely, infants with d-transposition of the great arteries and an intact ventricular septum normally present with significant cyanosis early after birth. This is secondary to decreased intercirculatory mixing, which typically requires early intervention to establish adequate intercirculatory mixing. It should be noted, however, that a large patent arterial duct in this cohort of patients may confuse the clinical picture as increased pulmonary blood flow in this intercirculatory shunt will blunt the degree of cyanosis and mimic the clinical course of those with increased pulmonary blood flow. An example of this case and intentional restriction of the arterial duct was reported by Masutani et al.[9] In d-transposition of the great arteries with obstruction of the left ventricular outflow tract obstruction – restricted effective pulmonary blood flow – the degree of cyanosis can be amplified and may not be affected by the degree of intercirculatory mixing due to a fixed restriction of pulmonary blood flow. If right ventricular outflow tract obstruction – restricted effective systemic blood flow – is present, pulmonary blood flow and oxygenation may be adequate but the infant will suffer from systemic hypoperfusion.

Pre-operative management

The pre-operative management of d-transposition of the great arteries focuses on ensuring adequate systemic perfusion and oxygen delivery that is dependent on adequate mixing of the systemic and pulmonary circulations. Depending on the initial presentation of the infant, all emergent resuscitative measures should be addressed immediately following standard neonatal advanced life support algorithms. Umbilical arterial and venous lines may be helpful for close surveillance of haemodynamics, acid–base status, and medication administration. Following stabilisation, prompt, directed cardiac evaluation starting with echocardiography should be initiated, especially for the cyanotic infant. Accordingly, in cases in which a prenatal diagnosis is available, it is preferable to deliver these infants in or near a surgical referral centre.[10]

Initiation of prostaglandin E1 may temporarily improve severe cyanosis by improving intercirculatory mixing – effective systemic and pulmonary blood flow – at the level of the arterial duct. Infusion of prostaglandin E1 can be initiated immediately after birth while awaiting completion of diagnostic studies or during transfer to a surgical centre. Once a diagnosis of d-transposition of the great arteries is confirmed, the prostaglandin E1 infusion can be discontinued if there is adequate mixing at the atrial or ventricular level. In the case of intact ventricular septum, prostaglandin E1 is usually infused until balloon atrial septostomy, or, in some cases, until surgery. Factors predictive for the successful discontinuation of prostaglandin E1 after balloon atrial septostomy in infants with intact ventricular septum have yet to be determined.[11] Pre-operative duration of prostaglandin E1 infusion has not been associated with adverse effects in post-operative outcomes.[12]

Risks associated with prostaglandin E1 include apnoea, fever, and vasodilation (hypotension). In addition, long-term exposure to prostaglandin therapy has been associated with an increased incidence of pyloric stenosis. Although the risk of apnoea is of significant consequence, the administration of aminophylline or caffeine may ameliorate this risk.[13] Elective intubation for infants being transported on prostaglandin E1 has been shown to be associated with an increased risk of major transport complications that outweigh any potential benefit.[14] In addition, shunting across the patent arterial duct may adversely increase pulmonary blood flow and cause symptoms related to pulmonary congestion. It should be noted, however, that mixing at the patent arterial duct alone may not be sufficient and that continued cyanosis may persist in the presence of a restrictive atrial septal defect.

Restriction to mixing at the atrial septum, particularly in infants with a patent arterial duct, may require prompt balloon atrial septostomy to relieve profound cyanosis until definitive surgical

treatment can be performed. Recently, Punn and Silverman[15] found that prenatal echocardiographic findings of a hypermobile atrial septum and reverse diastolic patent arterial duct flow were predictive of the need for balloon atrial septostomy. This percutaneous procedure may be performed under echocardiographic guidance at the bedside in an intensive care unit or under fluoroscopy in a cardiac catheterisation laboratory.[16,17] The safety of balloon atrial septostomy with regard to brain injury has been debated. McQuillen et al[18] published data on a series of infants with increased rates of brain injury associated with balloon septostomy. However, more recent data have shown balloon atrial septostomy to be safe and not related to pre-operative brain injury.[19–21] In fact, in the series by Petit et al[19], failure to achieve a pre-operative arterial oxygen content >40 mmHg was associated with brain injury. Although balloon atrial septostomy may be required in centres that cannot perform an immediate arterial switch operation, there are new data reporting successful arterial switch operation in the first few hours of life in infants, including those with intact ventricular septum and restrictive patent oval foramen.[22,23]

Changes in pulmonary vasculature may result in persistent primary pulmonary hypertension of the newborn in infants with d-transposition of the great arteries. This finding is more frequent in uncorrected, older infants with advanced pulmonary disease, but may occur earlier in those with intact ventricular septum and a patent arterial duct.[24] The development of pulmonary vascular disease is most likely multi-factorial, resulting from increased pulmonary blood flow, pressure, and hypoxia. Mortality is high in this group of infants and mid-term post-operative outcomes are adversely affected.[25,26] A number of medical management strategies have been proposed including the use inhaled nitric oxide[27] and bosentan.[28] Successful surgical management utilising inhaled nitric oxide in conjunction with extracorporeal membrane oxygenation pre-operatively has also been reported.[29,30]

Post-operative management

Several anatomic and physiologic factors influence the post-operative management and intensive care unit course after the arterial switch operation for d-transposition of the great arteries. Such factors may include low cardiac output syndrome, abnormal left ventricular compliance, mitral regurgitation, left atrial hypertension, and coronary ischaemia.

Post-operative low cardiac output syndrome was classically described in this patient population[31] and can be due to a number of causes after the arterial switch operation. Similar to other cardiac operations requiring cardiopulmonary bypass and circulatory arrest, suboptimal myocardial preservation, mechanical disruption of the myocardium, and the post-inflammatory effects of bypass should be considered. Unique to the infant with d-transposition of the great arteries, especially in the setting of an intact ventricular septum and delayed surgical intervention, is an untrained left ventricle that must now perform against systemic afterload. This predisposes the infant to declining left ventricular function, which may result in increasing mitral regurgitation. Ultimately, increased left atrial pressure develops, putting the infant at risk for increased pulmonary venous pressure. Standard monitoring including urine output, heart rate and blood pressure trends, mixed venous saturation, and serial lactate measurement can help alert the practitioner to declining cardiac output.

Management of low cardiac output during the post-operative period varies depending upon the physiologic state of the infant. Successful management is often achieved by developing a low-pressure, high-flow state as the left ventricle adapts to the demands of systemic afterload. Inotropic agents such as milrinone, epinephrine, or calcium can be titrated accordingly to help improve cardiac output while systemic afterload reduction can be achieved with alpha blockade – phenoxybenzamine or phentolamine – milrinone, and nitroprusside. During resuscitation, volume must be used with caution owing to the possibility of left atrial distension, high left atrial pressure, mitral regurgitation, and ventricular dysfunction. In situations where adequate haemodynamics cannot be achieved, a brief period of extracorporeal membrane oxygenation may be required to allow for ventricular recovery or surgical revision of any residual anatomic defects. Lastly, the pre-operative administration of steroids has been shown to mitigate the inflammatory effects of cardiopulmonary bypass while providing myocardial protection.[32,33]

A vital component of the atrial switch operation includes implantation of the coronary arteries into the neo-aorta. Therefore, vigilant monitoring and heightened awareness for signs of coronary insufficiency is of utmost importance. An immediate post-operative electrocardiogram should be obtained and compared against a pre-operative electrocardiogram. In the event of coronary insufficiency, nitroglycerin may be used; however, echocardiography and/or coronary angiography should be performed immediately to better delineate coronary anatomy, as surgical revision may be required.

Common arterial trunk

Collett and Edwards[34] initially described common arterial trunk in 1949 and classified the defect on the basis of the origin of the pulmonary arteries from the common arterial trunk. In 1965, Van Praagh and Van Praagh[35] revised the classification to be based on the presence or absence of the conotruncal septum. In assessing the type of common arterial trunk, thorough evaluation of the semilunar truncal valve, patency of the branch pulmonary arteries, presence of a patent arterial duct, and characteristics of the ventricular septal defect are all necessary for surgical planning and medical management pre-operatively. The truncal valve can have multiple leaflets leading to truncal valve insufficiency. Patients who required concomitant truncal valve surgery have been shown to have a mortality of 30% compared with 10% for those with isolated common arterial trunk repair.[36]

There are several associated cardiac defects that need to be identified in patients with common arterial trunk pre-operatively. The most common include anomalies of the coronary arteries (~15%), interrupted aortic arch, right aortic arch, patent arterial duct, persistent left superior caval vein, atrial septal defect, and an anomalous subclavian artery.[37,38] Identifying these defects early is imperative to reducing mortality and addressing the approach to surgical correction. Certain associated defects, such as interrupted aortic arch, have been shown to significantly increase mortality.[39] Non-cardiac anomalies and common cardiac-associated genetic syndromes, such as 22q11.2 deletion syndromes, should be ruled out before the patient undergoes surgical repair. It has been reported that up to 30% of patients with common arterial trunk have 22q11.2 deletion.[40–42]

Pre-operative management

Patients with common arterial trunk have functionally univentricular physiology and typically present clinically on the basis of the degree of pulmonary blood flow restriction. Patients presenting with cyanosis will likely have increased resistance in the pulmonary circulation secondary to pulmonary artery stenosis, pulmonary artery hypoplasia, or persistently elevated pulmonary vascular resistance. Patients with well-balanced pulmonary-to-systemic blood flow will likely not present until pulmonary vascular resistance falls. Those patients will then present with cardiac failure symptoms secondary to left ventricular volume overload and resultant pulmonary vascular congestion. Neonates who present in early cardiac failure or cardiogenic shock may have severe truncal valve stenosis or insufficiency.[36,37] Common arterial trunk has been independently associated with necrotising enterocolitis in term neonates with congenital heart disease admitted to the cardiac intensive care unit.[43] This is thought to be secondary to relative ischaemia of the gut due to excessive pulmonary blood flow resulting in insufficient systemic blood flow. Feeding intolerance and abdominal distension should be closely monitored for and promptly responded to with a low threshold to discontinue feedings. Neonates that develop necrotising enterocolitis but are identified early may not have increased mortality; however, it has been shown to significantly increase their hospital stay.[43]

Management of patients with 22q11.2 deletion syndrome in the critical care setting includes recognition of potential airway issues, feeding intolerance, immunodeficiency, and electrolyte abnormalities. Endotracheal intubation in these patients may be potentially difficult given their relative microstomia and retrognathia. In addition, laryngotracheomalacia may be present in these infants, and as surgery typically occurs at an early post-natal age this may complicate extubation. Vascular rings formed by a right aortic arch and aberrant left subclavian artery with left-sided ligamentum arteriosum can present with stridor and wheezing and should be assessed on pre-operative non-invasive imaging.[44] Feeding issues secondary to palatoschisis – cleft palate – and velopharyngeal incompetence should be anticipated in this patient population and an age-appropriate feeding plan should be formulated early. Given the increased risk of infectious complications secondary to T-cell immunodeficiency and hypogammaglobulinaemia, it is important that these patients receive irradiated blood products.[45] In addition, immunologic evaluation by a Paediatric Allergy/Immunology specialist may be necessary in the peri-operative period as prophylactic antibiotics may be warranted. Knowledge of additional associated anomalies of 22q11.2 deletion syndrome, including renal anomalies, anal atresia, and tethered cord syndrome, is also important.

Post-operative management

Post-operative physiology of common arterial trunk depends on the extent of the repair and the degree of residual lesions. Post-operative management must also account for the detrimental effects of cardiopulmonary bypass resulting in alterations of pulmonary and systemic vasculature resistance and myocardial dysfunction. Low cardiac output syndrome following cardiopulmonary bypass is common and of significant concern.[46] Depressed myocardial function following cardiopulmonary

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bypass is commonly treated with vasoactive infusion support. Contributing to the difficulty in post-operative management is right ventricular diastolic dysfunction. This can result from cardiopulmonary bypass-induced injury, ventriculotomy-related injury, or elevated pulmonary arterial pressure, which results in insufficient preload of the right ventricle. In the absence of an atrial communication, the patient will subsequently develop low cardiac output owing to insufficient preloading of the left ventricle. Placement of a left atrial line to allow closer monitoring for pulmonary arterial hypertension and associated crises should be considered, particularly in patients with higher risk for morality. Agitation secondary to pain may trigger a life-threatening pulmonary hypertensive crisis; therefore, it is imperative to maintain adequate sedation and paralysis for the first 24 to 48 hours after repair. Maintaining a relative metabolic alkalosis and keeping the patient relatively normocapnic will lead to pulmonary arterial bed vasodilation and decreased pulmonary vascular resistance. In addition, inhaled nitric oxide can be used to decrease pulmonary vascular resistance, and delayed sternal closure can be used when needed until pulmonary vascular resistance recovers. Extra-corporeal membrane oxygenation may be necessary for significant low cardiac output syndrome and/or pulmonary hypertension.

The most likely complication in critically ill neonates with 22q11.2 deletion syndrome is hypocalcaemia. Post-operative hypocalcaemia has been reported to occur in as high as 86% of patients with 22q11.2 deletion syndrome compared with 47% of patients without the deletion. A comparison of the two groups of patients with 22q11.2 deletion showed that those with post-operative hypocalcaemia were more likely to have significant complications and mortality.[47] In addition, neonatal myocardium is more dependent on exogenous calcium because of an immature sarcoplasmic reticulum. Therefore, hypocalcaemia will have a greater effect on cardiac function, cardiac rhythm, and vasomotor tone in these patients, and more frequent monitoring of ionised calcium should be performed in post-operative neonates with 22q11.2 deletion.

Conclusion

Although management of patients with unconventional physiologies such as those with d-transposition of the great arteries and common arterial trunk remains challenging, advances in prenatal diagnosis and pre- and post-operative management have significantly improved survival in these populations. There remains significant room for improvement, particularly with regard to morbidity. Future studies should focus on optimising and standardising in- and out-of-hospital care, with a goal of improving morbidity, mortality, and functional outcomes.

References

1. Marek J, Tomek V, Skovranek J, et al. Prenatal ultrasound screening of congenital heart disease in an unselected national population: a 21-year experience. Heart 2011; 97: 124–130.
2. Ferencz C, Rubin JD, McCarter RJ, et al. Congenital heart disease: prevalence at livebirth. The Baltimore-Washington Infant Study. Am J Epidemiol 1985; 121: 31–36.
3. Liebman J, Cullum L, Belloc NB. Natural history of transposition of the great arteries. Anatomy and birth and death characteristics. Circulation 1969; 40: 237–262.
4. Lim HG, Kim WH, Lee JR, Kim YJ. Long-term results of the arterial switch operation for ventriculo-arterial discordance. Eur J Cardiothorac Surg 2012, May 9 [epub ahead of print].
5. Fricke TA, d'Udekem Y, Richardson M, et al. Outcomes of the arterial switch operation for transposition of the great arteries: 25 years of experience. Ann Thorac Surg 2012; 94: 139–145.
6. Kempny A, Wustmann K, Borgia F, et al. Outcome in adult patients after arterial switch operation for transposition of the great arteries. Int J Cardiol 2012, Aug 9 [epub ahead of print].
7. Oda S, Nakano T, Sugiura J, et al. Twenty-eight years' experience of arterial switch operation for transposition of the great arteries in a single institution. Eur J Cardiothorac Surg 2012; 42: 674–679.
8. Wernovsky G. Transposition of the great arteries. In: Allen HD, Shaddy R, Feltes T (eds). Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. Wolters Kluwer Health/Lippincott Williams & Wilkins, Philadelphia, 2008, 1027–1084.
9. Masutani S, Seki M, Taketazu M, Senzaki H. Successful management of the persistent pulmonary hypertension of the newborn with transposition of the great arteries by restricted patency of the ductus arteriosus: a simple and rational novel strategy. Pediatr Cardiol 2009; 30: 1003–1005.
10. Hellstrom-Westas L, Hanseus K, Jogi P, et al. Long-distance transports of newborn infants with congenital heart disease. Pediatr Cardiol 2001; 22: 380–384.
11. Oxenius A, Hug MI, Dodge-Khatami A, et al. Do predictors exist for a successful withdrawal of preoperative prostaglandin E1 from neonates with d-transposition of the great arteries and intact ventricular septum? Pediatr Cardiol 2010; 31: 1198–1202.
12. Butts RJ, Ellis AR, Bradley SM, et al. Effect of prostaglandin duration on outcomes in transposition of the great arteries with intact ventricular septum. Congenit Heart Dis 2012; 7: 387–391.
13. Lim DS, Kulik TJ, Kim DW, et al. Aminophylline for the prevention of apnea during prostaglandin E1 infusion. Pediatrics 2003; 112 (1 Pt 1): e27–e29.
14. Meckler GD, Lowe C. To intubate or not to intubate? Transporting infants on prostaglandin E1. Pediatrics 2009; 123: 8.
15. Punn R, Silverman NH. Fetal predictors of urgent balloon atrial septostomy in neonates with complete transposition. J Am Soc Echocardiogr 2011; 24: 425–430.
16. Martin AC, Rigby ML, Penny DJ, Redington AN. Bedside balloon atrial septostomy on neonatal units. Arch Dis Child Fetal Neonatal Ed 2003; 88: F339–F340.
17. Zellers TM, Dixon K, Moake L, et al. Bedside balloon atrial septostomy is safe, efficacious, and cost-effective compared with septostomy performed in the cardiac catheterization laboratory. Am J Cardiol 2002; 89: 613–615.
18. McQuillen PS, Hamrick SE, Perez MJ, et al. Balloon atrial septostomy is associated with preoperative stroke in neonates with transposition of the great arteries. Circulation 2006; 113: 280–285.

19. Petit CJ, Rome JJ, Wernovsky G, et al. Preoperative brain injury in transposition of the great arteries is associated with oxygenation and time to surgery, not balloon atrial septostomy. Circulation 2009; 119: 709–716.
20. Beca J, Gunn J, Coleman L, et al. Pre-operative brain injury in newborn infants with transposition of the great arteries occurs at rates similar to other complex congenital heart disease and is not related to balloon atrial septostomy. J Am Coll Cardiol 2009; 53: 1807–1811.
21. Applegate SE, Lim DS. Incidence of stroke in patients with d-transposition of the great arteries that undergo balloon atrial septostomy in the University Healthsystem Consortium Clinical Data Base/Resource Manager. Catheter Cardiovasc Interv 2010; 76: 129–131.
22. Nevvazhay T, Chernogrivov A, Biryukov E, et al. Arterial switch in the first hours of life: no need for Rashkind septostomy? Eur J Cardiothorac Surg 2012; 42: 520–523.
23. Chasovskyi K, Fedevych O, Vorobiova G, et al. Arterial switch operation in the first hours of life using autologous umbilical cord blood. Ann Thorac Surg 2012; 93: 1571–1576.
24. Newfeld EA, Paul MM, Muster AJ, Idriss FS. Pulmonary vascular disease in complete transposition of the great arteries: a study of 200 patients. Am J Cardiol 1974; 34: 75–82.
25. Fan H, Hu S, Zheng Z, et al. Do patients with complete transposition of the great arteries and severe pulmonary hypertension benefit from an arterial switch operation? Ann Thorac Surg 2011; 91: 181–186.
26. Roofthooft MT, Bergman KA, Waterbolk TW, et al. Persistent pulmonary hypertension of the newborn with transposition of the great arteries. Ann Thorac Surg 2007; 83: 1446–1450.
27. El-Segaier M, Hellstrom-Westas L, Wettrell G. Nitric oxide in neonatal transposition of the great arteries. Acta Paediatr 2005; 94: 912–916.
28. Goissen C, Ghyselen L, Tourneux P, et al. Persistent pulmonary hypertension of the newborn with transposition of the great arteries: successful treatment with bosentan. Eur J Pediatr 2008; 167: 437–440.
29. Jaillard S, Belli E, Rakza T, et al. Preoperative ECMO in transposition of the great arteries with persistent pulmonary hypertension. Ann Thorac Surg 2005; 79: 2155–2158.
30. Luciani GB, Chang AC, Starnes VA. Surgical repair of transposition of the great arteries in neonates with persistent pulmonary hypertension. Ann Thorac Surg 1996; 61: 800–805.
31. Wernovsky G, Wypij D, Jonas RA, et al. Postoperative course and hemodynamic profile after the arterial switch operation in neonates and infants. A comparison of low-flow cardiopulmonary bypass and circulatory arrest. Circulation 1995; 92: 2226–2235.
32. Heying R, Wehage E, Schumacher K, et al. Dexamethasone pretreatment provides antiinflammatory and myocardial protection in neonatal arterial switch operation. Ann Thorac Surg 2012; 93: 869–876.
33. Schroeder VA, Pearl JM, Schwartz SM, et al. Combined steroid treatment for congenital heart surgery improves oxygen delivery and reduces postbypass inflammatory mediator expression. Circulation 2003; 107: 2823–2828.
34. Collett RW, Edwards JE. Persistent truncus arteriosus: a classification according to anatomic types. Surg Clin North Am 1949; 29: 1245–1270.
35. Van Praagh R, Van Praagh S. Anatomy of common aorticopulmonary trunk (truncus arteriosus communis) and its embryologic implications – a study of 57 necropsy cases. Am J Cardiol 1965; 16: 406–407.
36. Russell HM, Pasquali SK, Jacobs JP, et al. Outcomes of repair of common arterial trunk with truncal valve surgery: a review of the society of thoracic surgeons congenital heart surgery database. Ann Thorac Surg 2012; 93: 164–169.
37. Elzein C, Ilbawi M, Kumar S, et al. Severe truncal valve stenosis: diagnosis and management. J Card Surg 2005; 20: 589–593.
38. Tlaskal T, Hucin B, Kucera V, et al. Repair of persistent truncus arteriosus with interrupted aortic arch. Eur J Cardiothorac Surg 2005; 28: 736–741.
39. Bohuta L, Hussein A, Fricke TA, et al. Surgical repair of truncus arteriosus associated with interrupted aortic arch: long-term outcomes. Ann Thorac Surg 2011; 91: 1473–1477.
40. Goldmuntz E, Emanuel BS. Genetic disorders of cardiac morphogenesis. The DiGeorge and velocardiofacial syndromes. Circ Res 1997; 80: 437–443.
41. Carotti A, Digilio MC, Piacentini G, et al. Cardiac defects and results of cardiac surgery in 22q11.2 deletion syndrome. Dev Disabil Res Rev 2008; 14: 35–42.
42. Ryan AK, Goodship JA, Wilson DI, et al. Spectrum of clinical features associated with interstitial chromosome 22q11 deletions: a European collaborative study. J Med Genet 1997; 34: 798–804.
43. McElhinney DB, Hedrick HL, Bush DM, et al. Necrotizing enterocolitis in neonates with congenital heart disease: risk factors and outcomes. Pediatrics 2000; 106: 1080–1087.
44. McElhinney DB, McDonald-McGinn D, Zackai EH, et al. Cardiovascular anomalies in patients diagnosed with a chromosome 22q11 deletion beyond 6 months of age. Pediatrics 2001; 108: E104.
45. McDonald R, Dodgen A, Goyal S, et al. Impact of 22q11.2 deletion on the postoperative course of children after cardiac surgery. Pediatr Cardiol 2012, Aug 3 [epub ahead of print].
46. Hoffman TH, Wernovsky G, Atz AM, et al. Efficacy and safety of milrinone in preventing low cardiac output syndrome in infants and children after corrective surgery for congenital heart disease. Circulation 2003; 107: 996–1002.
47. Shen L, Gu H, Wang D, et al. Influence of chromosome 22q11.2 microdeletion on postoperative calcium level after cardiac-correction surgery. Pediatr Cardiol 2011; 32: 904–909.