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The evaluation of cases with double-inlet left ventricle-ventriculoarterial discordance

Çift girişli sol ventrikül-ventriküloarteriyel diskordans olgularının değerlendirilmesi

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ABSTRACT

Background: This study aims to evaluate the cases of functional univentricle with the diagnosis of double-inlet left ventricle-ventriculoarterial discordance.

Methods: Between January 2010 and October 2014, a total of 34 patients (17 males, 17 females; median age 2.64 months; range 1 day to 24 years) with double-inlet left ventricle-ventriculoarterial discordance were analyzed retrospectively. The demographic characteristics of the patients, echocardiographic and hemodynamic measurements, invasive, surgical procedures performed and their outcomes were reviewed.

Results: Of the patients, 24 had pulmonary hypertension with arch obstruction in eight of them. Eight had pulmonary stenosis, and two had pulmonary atresia. None of the patients had outflow obstruction as assessed by echocardiography. Surgery was performed in 25 patients. Of these, 14 patients underwent initial pulmonary artery banding procedure and four of these also underwent arch reconstruction. Bidirectional cavopulmonary connection was performed in six patients, aortopulmonary shunt in four patients, and the Norwood-type operation in one patient. The median follow-up was 9.96 months (range 0.24 to 53.88 months). During follow-up, 12 patients underwent a bidirectional cavopulmonary connection. Systemic outflow restriction did not develop in any of the patients who were initially palliated with pulmonary artery banding with or without arch reconstruction and proceeded to the bidirectional cavopulmonary shunt stage.

Conclusion: Based on our study findings, although the primary treatment strategy in patients with double-inlet left ventricle-ventriculoarterial discordance displays a great variability, aortopulmonary shunting or ductal stenting can be performed in patients with restricted pulmonary blood flow, pulmonary artery banding with or without arch reconstruction in patients with unrestricted pulmonary blood flow. The Norwood type operation can be performed initially in patients with bulboventricular foramen restriction.

Keywords: Childhood; double-inlet ventricle; ventriculoarterial discordance.

ÖZ

Amaç: Bu çalışmada çift girişli sol ventrikül-ventriküloarteriyel diskordans tanılı fonksiyonel tek ventriküllü olgular değerlendirildi.

Çalışma planı: Ocak 2010 - Ekim 2014 tarihleri arasında çift girişli sol ventrikül-ventriküloarteriyel diskordanslı toplam 34 hasta (17 erkek, 17 kadın; ort. yaş 2.64 ay; dağılım 1 gün-24 yıl) retrospektif olarak incelendi. Hastaların demografik özellikleri, ekokardiyografik ve hemodinamik ölçümleri, yapılan invazif, cerrahi işlemler ve sonuçları gözden geçirildi.

Bulgular: Hastaların 24'ünde pulmoner hipertansiyon olup, bunların sekizinde arkusta tıkanıklık vardı. Sekizinde pulmoner darlık ve ikisinde pulmoner atrezi mevcuttu. Hiçbir hastada ekokardiyografi ile değerlendirildiği üzere, çıkım yolu tıkanıklığı saptanmadı. Yirmi beş hastaya cerrahi yapıldı. Bu hastaların 14'üne pulmoner arter bant işlemi ve bunların dördüne ek olarak arkus rekonstrüksiyonu gerçekleştirildi. Altı hastaya iki yönlü kavopulmoner anastomoz, dört hastaya aortopulmoner şant ve bir hastaya Norwood tipi cerrahi yapıldı. Medyan takip süresi 9.96 ay (dağılım 0.24-53.88 ay) idi. Takip sırasında 12 hastaya iki yönlü kavopulmoner anastomoz yapıldı. İlk palyasyon olarak arkus rekonstrüksiyonu ile birlikte veya tek başına pulmoner arter bant yapılan ve iki yönlü kavopulmoner şant dönemine gelen hiçbir hastada çıkım yolunda sistemik darlık gelişmedi.

Sonuç: Çalışma bulgularımıza göre, çift girişli sol ventrikül ventriküloarteriyel diskordanslı hastalarda birincil tedavi stratejisi büyük farklılıklar göstermekle birlikte, pulmoner kan akımı sınırlı olan hastalara aortopulmoner şant veya duktal stent uygulanabilirken, pulmoner kan akımı sınırlı olmayan hastalara arkus rekonstrüksiyonu ile beraber veya tek başına pulmoner arter bandı uygulanabilir. Bulboventriküler foramen restriksiyonu olan hastalara ise ilk olarak Norwood tipi cerrahi uygulanabilir.

Anahtar sözcükler: Çocukluk çağı; çift girişli ventrikül; ventriküloarteriyel diskordans.

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Double-inlet left ventricle (DILV) is the most common form of the anatomic univentricular heart.[1,2] A discordant ventriculoarterial connection is mostly seen in the setting of a DILV with hypoplastic right ventricle on the left side; however, a concordant ventriculoarterial connection or a double-outlet from the right or left ventricle can be also seen.[2-6] Pulmonary hypertension with aortic coarctation or the interruption of the aorta often presents with a pathology in which the systemic outflow traverses a bulboventricular foramen, which has a propensity to narrow over time. The restriction of the bulboventricular foramen and subaortic obstruction is major complications in these patients.[7-10]

Herein, we aimed to evaluate the patients with functional univentricular heart with the diagnosis of DILV- ventriculoarterial discordance (VAD).

PATIENTS AND METHODS

In this study, we retrospectively evaluated a total of 34 patients (17 males, 17 females; median age 2.64 months; range 1 day to 24 years) with DILV-VAD between January 2010 and September 2014.

The echocardiographic evaluations, catheter studies, and surgical reports were reviewed retrospectively from the patient records. The surgery types (aortopulmonary shunt, pulmonary artery banding with or without arch reconstruction, bidirectional cavopulmonary connection, etc.), timing of procedures, and outcomes were noted.

The successive echocardiographic measurements of the bulboventricular foramen/aortic annulus ratio, the bulboventricular foramen area index, and the gradient across the bulboventricular foramen were recorded in the regularly followed patients. Hypoplasia of the aortic arch and isthmus, coarctation and associated cardiac findings were also recorded.

Systemic outflow obstruction was defined as a resting peak instantaneous gradient higher than 20 mmHg on echocardiography or a peak-to-peak gradient higher than 10 mmHg with cardiac catheterization.[7,8] The bulboventricular foramen was considered restricted, if the ratio of the bulboventricular foramen diameter to the aortic annulus diameter was ≤0.5 or tended to be restricted, if the bulboventricular foramen diameter to the aortic annulus diameter was smaller than 1 or the bulboventricular foramen area index was smaller than 2 cm^2/m^2.[9,10]

Aortopulmonary shunt procedures were performed by median sternotomy. Pulmonary artery banding procedures were performed by superior mini sternotomy, left anterior thoracotomy, or median sternotomy in patients requiring (arch reconstruction, particularly. Pulmonary artery banding was performed by fixing 3 mm thick silicone sheath to the opposite sites of the pulmonary artery. Arch reconstruction in patients with low arch Z scores (under -2) and coarctation repair in case of a gradient higher than 20 mmHg with diastolic continuity were also performed. During the operation, the band was tightened up to a level where pulmonary pressure was 50% of systemic pressure with the room-air oxygen saturation around 75%, while keeping the near-infrared spectroscopy (NIRS) and hematocrit level above 40% and central venous pressure around 10 mmHg.

Bidirectional cavopulmonary connection procedure was performed after six months by resternotomy. Some patients were operated on-pump, while some of them off-pump. We performed routine catheterization and evaluated pulmonary artery pressures before bidirectional cavopulmonary connection procedure. In patients with elevated pulmonary artery pressure, particularly, in patients who had previously pulmonary artery banding, bidirectional cavopulmonary connection was postponed and the catheterization procedure was repeated after medical treatment or tightening of the pulmonary band.

The study protocol was approved by the institutional Ethics Committee. The study was conducted in accordance with the principles of the Helsinki Declaration.

Statistical analysis

Statistical analysis was performed using the SPSS version 15.0 for Windows software (SPSS Inc., Chicago, IL, USA). Data were presented in median (minimum-maximum) or mean ± standard deviation. Statistical significances of continuous variables were determined by using non-parametric Mann-Whitney U test, while categorical variables were analyzed by the Fisher's exact test. A p value of <0.05 was considered statistically significant.

RESULTS

Abdominal and atrial situs were solitus in all of the patients. The position of the heart was levocardia in all, but one patient with mesocardia. Ventricular looping was d-loop (right-handed) in 19 patients (55.9%) and l-loop (left-handed) in 15 patients (44.1%).

Twenty-four patients had pulmonary hypertension, eight had pulmonary stenosis, and two had pulmonary atresia.

The sequential bulboventricular foramen diameter/aortic annulus diameter ratio and gradient across the bulboventricular foramen were recorded in regularly followed-up patients. The mean initial bulboventricular foramen/aortic annulus ratio was 0.9±0.2 (0.64-1.2), and the mean initial bulboventricular foramen area index was 1.71±0.88 cm^2/m^2 (0.57-3.41). The mean bulboventricular foramen/aortic annulus ratio was 0.91±0.35 (0.55-1.68), and the mean bulboventricular foramen area index was 2.29±1.37 cm^2/m^2 (1.03-4.79) during follow-up. There were no statistically significant differences between the initial and follow-up bulboventricular foramen/aortic annulus ratios and bulboventricular foramen area index values (p=0.74, p=0.71, respectively).

None of the patients had a bulboventricular foramen/annulus ratio of ≤0.5 on admission or before any procedure. Similarly, none of the bulboventricular foramen gradients were more than 20 mmHg on admission or before any procedure.

Hemodynamic studies were performed in 16 patients, twice in four of the patients. Ductal stenting was performed in two patients; one with severe pulmonary stenosis. The stent was removed during the bidirectional cavopulmonary shunting. The other patient had pulmonary atresia; the stent was obstructed for a short time period and an aortopulmonary shunt was performed. A persistent left superior vena cava occlusion with device at the drainage site to the coronary sinus was performed in one patient, before the bidirectional cavopulmonary shunting. A balloon angioplasty for coarctation was performed in one patient, and vasoreactivity was tested in four. Vasoreactivity was also tested in two patients without pulmonary stenosis who were diagnosed in the late stage and in a patient with pulmonary artery banding which would likely to be ineffective. After cathether studies and vasoreactivity tests, these patients were accepted as ineligible for surgery; therefore, medical treatment was started. In addition, in a patient who underwent persistent left superior vena cava occlusion, a test occlusion with balloon and a vasoreactivity test were performed to show the changes in pulmonary artery pressure before occlusion.

Furthermore, 29 of 34 patients (85%) with DILV-VAD were followed on a regular basis with a median duration of 10.8 (range 2 to 53.88) months. Five patients residing in distant regions were lost during follow-up. Of these, three patients were lost after bidirectional cavopulmonary connection with or without pulmonary artery reconstruction, whereas two patients were lost after pulmonary artery banding. In addition, four patients died during follow-up. One of these died two years after a shunt procedure with a sudden hemodynamic deterioration and sepsis. Three patients who underwent different operations died during the early postoperative period in the intensive care unit or ward stay. One of the patients died at 10 months of age, two weeks after the bidirectional cavopulmonary connection operation, and another died at two months of age in the second postoperative day due to low cardiac output after pulmonary artery banding. Another patient with transverse arch hypoplasia and severe coarctation died at five months of age due to sepsis before discharge, after the Norwood-type operation.

Of the patients with DILV-VAD, 73.5% (n=25) were operated immediately after the initial diagnosis or during follow-up (Figure 1). Among 25 operated patients, an initial shunt operation was performed in four, pulmonary artery banding in 14, bidirectional cavopulmonary connection in six, and Norwood-type operation in one. The median age of initial palliation was two (range 0.33 to 96) months.

Aortopulmonary shunting was performed in three patients with severe pulmonary stenosis and one patient with pulmonary atresia during the first month of age. A Blalock Taussig-type systemic-pulmonary shunt was performed to supply pulmonary blood flow in three patients. In the last patient with pulmonary atresia, a central shunt was performed, when our attempt for ductal stenting failed. Subsequently, two patients with systemic-pulmonary shunt underwent a bidirectional cavopulmonary connection procedure.

Of the patients with DILV-VAD, eight had aortic arch and isthmus hypoplasia during the initial evaluation by echocardiography, which was also confirmed by computed tomography. Initial palliation with pulmonary artery banding was performed in conjuction with aortic arch reconstruction in four of these patients and together with main pulmonary artery reconstruction in one of these four patients. In addition, pulmonary artery banding without arch reconstruction was performed in another patient after perioperative re-evaluation of the arch and isthmus. Three of the eight patients were scheduled for surgery (i.e., pulmonary artery banding with arch reconstruction) and were kept in the waiting list.

A patient with severe transverse arch hypoplasia and coarctation who was late for pulmonary artery banding underwent the Norwood-type operation of

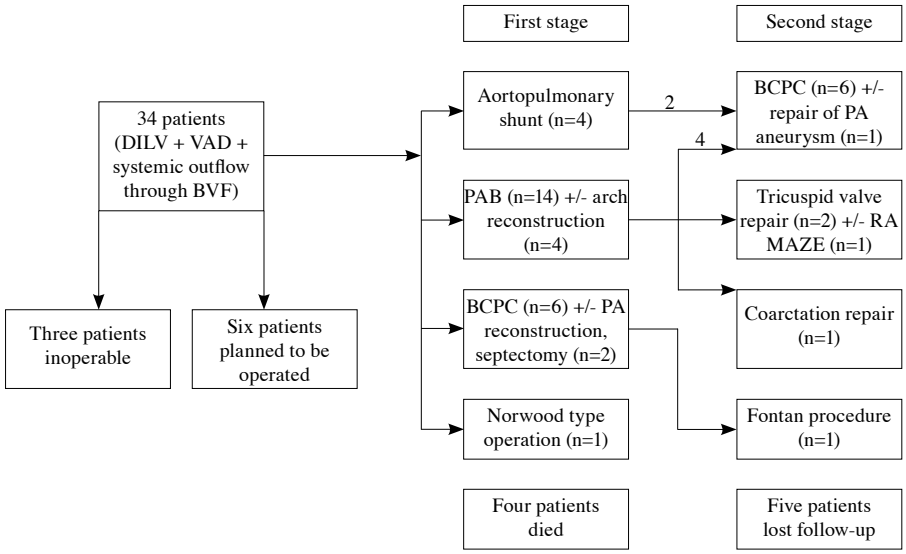

Figure 1. Outcome and surgical data of patients with double inlet left ventricle and ventriculoarterial discordance.

DILV: Double-inlet left ventricle; VAD: Ventriculoarterial discordance; BVF: Bulboventricular foramen; BCPC: Bidirectional cavopulmonary connection; PAB: Pulmonary artery banding; RA: Right atrium; MAZE: Heart Surgery for Atrial Fibrillation; PA: Pulmonary artery.

arch reconstruction with a right modified Blalock-Taussig shunt to relieve severe obstruction.

The median age of pulmonary artery banding in 14 patients was two (range 0.5 to 5) months. Seven of these patients who had pulmonary artery banding were reoperated and four of them had bidirectional cavopulmonary connection (Figure 1). Aortic coarctation developed in one of the patients five months after the pulmonary artery banding during follow-up and was repaired with an excision and end-to-end anastomosis of the coarcted segment. Two patients had tricuspid valve repair due to severe tricuspid regurgitation and one of them had an additional right atrial Maze procedure due to intractable arrhythmia.

Bidirectional cavopulmonary connection was completed in 12 patients during follow-up (bidirectional cavopulmonary connection was performed as initial operations in six patients). The median age at the time of the bidirectional cavopulmonary connection was 13 (range 8 to 156) months. The initial bidirectional cavopulmonary connection in one patient was completed to the Fontan circulation. Bulboventricular foramen restriction was not reported in any of the patients after the pulmonary artery banding procedure.

Surgery was planned for six patients. The planned procedures included pulmonary artery banding and arch reconstruction for three of them and bidirectional cavopulmonary connection for the other three patients.

DISCUSSION

Double-inlet ventricle in which a large ventricle, mostly with the morphology of a left ventricle, receives both atrioventricular valves and connects to a hypoplastic ventricle through a ventricular septal defect, is the most common form of the anatomic univentricular heart.[1] In a morphologic study with 54 autopsied specimens and 43 clinical DILV patients, Uemura et al.[11] reported that 64% had hypoplastic right ventricle leftwards, while 36% had hypoplastic right ventricle rightwards. They reported the most common stereotype was clinically known as the SLL (Situs solitus, L-looping ventricle, aorta is left anterior to pulmonary artery) type; however, 45.7% patients were SDD (Situs solitus, D-looping ventricle, aorta is right anterior to pulmonary artery) with the hypoplastic ventricle located anterosuperiorly and to the right. In a study by Frescura and Thiene,[12] of the hearts with DILV, the atrial situs was mostly solitus (in 29 of 35 cases) and the hypoplastic ventricle was located anterosuperiorly and to the right in most cases (in 23 of 29 cases). Consistent with these findings, in our study, the atrial situs was also solitus in all patients with DILV-VAD, while the hypoplastic ventricle was located anterior and to the right of the dominant morphological left ventricle (d-loop) in 55.9% of the patients.

Heart defects characterized by excessive pulmonary blood flow and hypoplasia of the aorta, such as DILV or tricuspid atresia with ventriculoarterial discordance, are pathologies in which systemic outflow traverses through a bulboventricular foramen which tends to narrow over time. The interventricular communication frequently causes subaortic stenosis in the stereotypical DILV with SLL.[9,10,13] Therefore, the main goal of initial palliation in patients DILV-VAD should be to relieve the arch obstruction and limit the pulmonary blood flow, which can be achieved by pulmonary artery banding with an arch reconstruction, or the Norwood-type operation.[14,15] Due to the heterogeneity of the anatomic substrates and the difficulty in identifying patients who would develop a foramen obstruction, there are different approaches for palliation of these patients.

In a study including 25 patients, Clarke et al.[16] evaluated the surgical outcomes of DILV and l-transposition morphology. The authors reported that they used arch repair and pulmonary artery banding as initial palliation to delay the timing of a Damus-Kaye-Stansel procedure or bulboventricular foramen resection, until the bidirectional cavopulmonary connection, which is the mainstay in the palliation of these patients.[2] Furthermore, they suggested that their approach compared favorably with reported results for the Norwood palliation.

The restriction of bulboventricular foramen in DILV or tricuspid atresia with a discordant ventriculoarterial connection is a condition recognized either at the time of the bidirectional cavopulmonary connection procedure or at the completion of the Fontan procedure.[17] It was suggested that an intervention to the ventricular outflow tract could have been applied earlier and prophylactically, even in those who did not eventually have the potential for developing a subaortic stenosis; however, this remains controversial.[8,11,18,19] Alsoufi et al.[20] reported the outcomes of a staged palliation in single ventricle patients who were at risk for systemic ventricular outflow tract obstruction. Of the patients, 18 had DILV-VAD. All patients underwent pulmonary artery banding with or without arch repair and atrial septectomy as the initial palliation, whereas the Damus-Kaye-Stansel operation was performed in all patients without any exception, even in those without an outflow gradient during the time of the bidirectional cavopulmonary connection operation. The authors suggested that pulmonary artery banding with or without arch repair and atrial septectomy as the initial palliation had several advantages, mainly avoiding a cardiopulmonary bypass in neonates, as well as several drawbacks, most importantly the possibility of ventricular hypertrophy which might accelerate the narrowing of bulboventricular foramen. In another study, Lotto et al.[14] evaluated the outcomes of the Norwood procedures in 24 patients with DILV-VAD who required arch repair. The authors reported the early mortality rate as 21.6% and 5-10 year survival rates as 72.8±7.4%. Ruzmetov et al.[21] evaluated the outcomes of initial pulmonary artery banding compared to the Norwood-type reconstruction in neonates. They reported that, although the Norwood-type reconstructions provided an unobstructed systemic outflow tract by avoiding the risk of heart block, freedom from any type of re-intervention and survival was similar with both procedures. Furthermore, in a case series including 15 patients, Odim et al.[22] reported that 47% patients did not develop subaortic obstruction following an initial pulmonary artery banding and progressed to a bidirectional cavopulmonary connection operation.

On the other hand, it may be challenging to determine, if a bulboventricular foramen is restricted or will be restricted, depending solely on the pressure gradient across the bulboventricular foramen, particularly in neonates with a patent ductus arteriosus. The bulboventricular foramen area index and bulboventricular foramen size, compared to the aortic annulus, are recommended to follow the foramen restriction.[9,10] In our study, no subaortic obstruction was detected on admission. The majority of the bulboventricular foramen area index values were smaller than 2 cm^2/m^2, although all but two of the bulboventricular foramen size/aortic annulus ratios were higher than 0.5 without a gradient across the bulboventricular foramen. This may suggest a high risk for the development of bulboventricular foramen restriction. Although all bulboventricular foramen size/aortic annulus ratios during follow-up were higher than 0.5 due to short follow-up period in our study, it would be rational to consider that patients who did not develop an obstruction might do so in the future; therefore, close follow-up of the subaortic area should be arranged for these patients.

In our clinic, pulmonary artery banding alone or with arch repair for patients with arch obstruction (unless severe atrioventricular regurgitation is present) and aortopulmonary shunting or ductal stenting or directly bidirectional cavopulmonary connection for patients with restriction of pulmonary blood flow were preferred as the initial palliation in patients with DILV-VAD. Patients were selected for arch repair

initially and followed by echocardiographic variables indicating bulboventricular foramen restriction rather than pulmonary artery banding with arch repair, followed by the Damus-Kaye-Stansel procedure at the time of bidirectional cavopulmonary connection operation for all patients, as depicted in the literature.[20] Only four of 14 patients with pulmonary artery banding had arch repair initially, while none of the patients who proceeded to the bidirectional cavopulmonary connection required for the Damus-Kaye-Stansel procedure or bulboventricular foramen resection, as further confirmed by echocardiography. However, the mean follow-up in our study was relatively short. As reported in the literature, bulboventricular foramen restriction is more frequent and develops much earlier in cases of an association with an arch pathology and systemic obstruction with gradient; these patients need to be followed closely for bulboventricular foramen restriction and ventricular hypertrophy, as it unlikely to influence the long-term outcomes without ventricular hypertrophy.[23-25] Arch pathologies should be also detailed by computed tomography imaging or angiographic procedures in conjuction with sequential measurements of echocardiographic Z scores.

Nonetheless, there are some limitations to this study. First, it has a retrospective design. Second, due to the rarity of these pathologies and as the Istanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Center is a relatively new center, our sample size was small with a short duration of follow-up. Third, as the surgical treatment strategies may vary depending on the surgeon, there is no specific therapy paradigm for these patient population.

In conclusion, patients with a functionally single ventricle, restricted or unrestricted pulmonary blood flow require staged palliation which serves preliminary steps to the Fontan operation. The treatment strategy in patients with double-inlet left ventricle with ventriculoarterial discordance displays great variability. Aortopulmonary shunt or ductal stenting can be performed in patients with restricted pulmonary blood flow, while the Norwood-type operation can be used initially in patients with unrestricted pulmonary blood flow and bulboventricular foramen restriction. In addition, pulmonary artery banding with or without arch reconstruction can be the procedure of choice in initial palliation in patients without systemic outflow obstruction with lower morbidity and mortality than in the Norwood-type operation. Also, bulboventricular foramen restriction which may develop over time in these patients can be handled by the Damus-Kaye-Stansel operation or bulboventricular foramen resection which will be performed during the bidirectional cavopulmonary shunt operation.

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REFERENCES

1. Wilkinson JL, Anderson RH. Anatomy of functionally single ventricle. World J Pediatr Congenit Heart Surg 2012;3:159-64.
2. Ekici B, Unsal H, Apaydın Z, Ekici E, İkizler C. The classic Glenn operation in the double inlet left ventricle: a 20-year follow-up. Turk Gogus Kalp Dama 2013;21:1023-6.
3. Anderson RH, Tynan M, Freedom RM, Quero-Jiménez M, Macartney FJ, Shinebourne EA, et al. Ventricular morphology in the univentricular heart. Herz 1979;4:184-97.
4. Doherty A, Ho SY, Anderson RH, Rigby ML. Morphological nature of the atrioventricular valves in hearts with double inlet left ventricle. Pediatr Pathol 1989;9:521-9.
5. Anderson RH, Lenox CC, Zuberbuhler JR, Ho SY, Smith A, Wilkinson JL. Double-inlet left ventricle with rudimentary right ventricle and ventriculoarterial concordance. Am J Cardiol 1983;52:573-7.
6. Penny DJ, Anderson RH. Double-inlet ventricle: morphology: double inlet left ventricle. In: Anderson RH, Baker EJ, Macartney FJ, Rigby ML, Shinebourne EA, Tynan M, editors. Paediatric Cardiology. Edinburgh: Harcourt Publishers Ltd; 2002. p. 1017-23.
7. Rychik J, Jacobs ML, Norwood WI Jr. Acute changes in left ventricular geometry after volume reduction operation. Ann Thorac Surg 1995;60:1267-73.
8. Karl TR, Watterson KG, Sano S, Mee RB. Operations for subaortic stenosis in univentricular hearts. Ann Thorac Surg 1991;52:420-7.
9. Freedom RM, Benson LN, Smallhorn JF, Williams WG, Trusler GA, Rowe RD. Subaortic stenosis, the univentricular heart, and banding of the pulmonary artery: an analysis of the courses of 43 patients with univentricular heart palliated by pulmonary artery banding. Circulation 1986;73:758-64.
10. Matitiau A, Geva T, Colan SD, Sluysmans T, Parness IA, Spevak PJ, et al. Bulboventricular foramen size in infants with double-inlet left ventricle or tricuspid atresia with transposed great arteries: influence on initial palliative operation and rate of growth. J Am Coll Cardiol 1992;19:142-8.
11. Uemura H, Ho SY, Adachi I, Yagihara T. Morphologic spectrum of ventriculoarterial connection in hearts with double inlet left ventricle: implications for surgical procedures. Ann Thorac Surg 2008;86:1321-7.
12. Frescura C, Thiene G. The new concept of univentricular heart. Front Pediatr 2014;2:62.
13. Franklin RC, Sullivan ID, Anderson RH, Shinebourne EA, Deanfield JE. Is banding of the pulmonary trunk obsolete for

infants with tricuspid atresia and double inlet ventricle with a discordant ventriculoarterial connection? Role of aortic arch obstruction and subaortic stenosis. J Am Coll Cardiol 1990;16:1455-64.

14. Lotto AA, Hosein R, Jones TJ, Barron DJ, Brawn WJ. Outcome of the Norwood procedure in the setting of transposition of the great arteries and functional single left ventricle. Eur J Cardiothorac Surg 2009;35:149-55.
15. Tchervenkov CI, Shum-Tim D, Béland MJ, Jutras L, Platt R. Single ventricle with systemic obstruction in early life: comparison of initial pulmonary artery banding versus the Norwood operation. Eur J Cardiothorac Surg 2001;19:671-7.
16. Clarke AJ, Kasahara S, Andrews DR, Cooper SG, Nicholson IA, Chard RB, et al. Mid-term results for double inlet left ventricle and similar morphologies: timing of Damus-Kaye-Stansel. Ann Thorac Surg 2004;78:650-7.
17. Lacour-Gayet F. Management of older single functioning ventricles with outlet obstruction due to a restricted "VSD" in double inlet left ventricle and in complex double outlet right ventricle. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2009:130-2.
18. Rychik J, Murdison KA, Chin AJ, Norwood WI. Surgical management of severe aortic outflow obstruction in lesions other than the hypoplastic left heart syndrome: use of a pulmonary artery to aorta anastomosis. J Am Coll Cardiol 1991;18:809-16.
19. van Son JA, Reddy VM, Haas GS, Hanley FL. Modified surgical techniques for relief of aortic obstruction in [S,L,L] hearts with rudimentary right ventricle and restrictive bulboventricular foramen. J Thorac Cardiovasc Surg 1995;110:909-15.
20. Alsoufi B, Al-Wadai A, Khan M, Al-Ahmadi M, Kalloghlian A, Bulbul Z, et al. Outcomes of Damus-Kaye-Stansel anastomosis at time of cavopulmonary connection in single ventricle patients at risk of developing systemic ventricular outflow tract obstruction. Eur J Cardiothorac Surg 2014;45:77-82.
21. Ruzmetov M, Geiss DM, Fortuna RS. Outcomes of double inlet left ventricle and similar morphologies: a single center comparison of initial pulmonary artery banding versus a norwood-type reconstruction. J Card Surg 2013;28:569-75.
22. Odim JN, Laks H, Drinkwater DC Jr, George BL, Yun J, Salem M, et al. Staged surgical approach to neonates with aortic obstruction and single-ventricle physiology. Ann Thorac Surg 1999;68:962-7.
23. Fiore AC, Rodefeld M, Vijay P, Turrentine M, Seithel C, Ruzmetov M, et al. Subaortic obstruction in univentricular heart: results using the double barrel Damus-Kaye Stansel operation. Eur J Cardiothorac Surg 2009;35:141-6.
24. Lan YT, Chang RK, Laks H. Outcome of patients with double-inlet left ventricle or tricuspid atresia with transposed great arteries. J Am Coll Cardiol 2004;43:113-9.
25. Ilbawi MN, DeLeon SY, Wilson WR Jr, Quinones JA, Roberson DA, Husayni TS, et al. Advantages of early relief of subaortic stenosis in single ventricle equivalents. Ann Thorac Surg 1991;52:842-9.