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Original Article

Longer-term issues for young adults with hypoplastic left heart syndrome: contraception, pregnancy, transition, transfer, counselling, and re-operation

Sameh M. Said,[1] Joseph A. Dearani,[1] Candice K. Silversides,[2] Richard M. Martinez,[3] David B. Drajpuch[4]

[1]Department of Cardiac Surgery, Mayo Clinic, Rochester, Minnesota; [2]Division of Cardiology, Mount Sinai and Toronto General Hospitals, Toronto, Ontario, Canada; [3]Division of Pediatric Cardiology, The Congenital Heart Institute of Florida (CHIF), All Children's Hospital, University of South Florida College of Medicine, Saint Petersburg, Florida; [4]Department of Cardiac Nursing, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, United States of America

Abstract Hypoplastic left heart syndrome remained a largely untreated lesion until the 1980s. In the current era, 75–80% of patients who are managed at "centres of excellence" can be expected to survive into young adulthood after staged palliation. This improved survival has led to an emerging population of patients now entering adulthood with a new set of concerns. We discuss the realised and potential issues that will be faced by these patients, including family planning, transition, and re-operation.

Keywords: Fontan; adult congenital heart disease; family planning; surgery

The incidence of congenital cardiac disease is eight per thousand newborns. Owing to the fact that outcomes have improved dramatically in the last four decades, at present over one million adults with congenital cardiac disease are alive in the United States of America, at least half of whom have complex congenital cardiac disease.[1]

In the Adult Congenital Heart Disease clinic of Tampa Bay, at least 10% of our patients have functionally univentricular physiology; however, our experience with adults with palliated hypoplastic left heart syndrome is only beginning.[2] Hypoplastic left heart syndrome remained a largely untreated lesion until the 1980s. In the current era, 75–80% of patients who are managed at "centres of excellence" can be expected to survive into young adulthood after staged palliation.[1]

Experience now exists of following up patients with functionally univentricular hearts into their 40s and, of late, early 50s. These patients have come to represent among our most challenging group of patients in terms of electrophysiological issues, as well as systolic and diastolic ventricular dysfunction. By contrast, most young adult patients with reconstructed hypoplastic left heart syndrome are now only in their second decade of life, and much is not known about their longer-term issues. One particular area of uncertainty is family planning.

Correspondence to: D. B. Drajpuch, MSN CRNP, Children's Hospital of Philadelphia, 12NW10, 34th Street and Civic Center Boulevard, Philadelphia, Pennsylvania 19104, United States of America. Tel: +1 215 590 2708; Fax: +1 215 590 2715; E-mail: drajpuchd@email.chop.edu

Contraception and pregnancy counselling

For women with hypoplastic left heart syndrome, contraception, and counselling about pregnancy should begin early to ensure safe and informed decisions. One must consider all of the following issues:

- safety and efficacy of contraception,
- risks of pregnancy,
- safety of cardiac medication during pregnancy, and
- the consequences of an unplanned pregnancy.

Input is often required from both a congenital cardiologist and a gynaecologist or obstetrician. Unfortunately, current practices regarding contraception and pregnancy are often suboptimal.[3]

Contraception

At present, very limited data exist about the risk of pregnancy, and no data exist about the safety of

contraception in women who have undergone staged reconstruction of hypoplastic left heart syndrome.[4] However, multiple studies exist about the risk of pregnancy in women with other underlying cardiac lesions, who have Fontan circulation,[5–8] and these data serve as a basis for many of the following recommendations. Knowledge of the risks of pregnancy will improve as more women with hypoplastic left heart syndrome reach childbearing age. It is therefore important to discuss these issues routinely to provide updated counselling.

Various forms of contraception are available, including the following methods:

- barrier methods – condoms or diaphragms,
- combined oestrogen and progestin contraceptives – oral pills, transdermal patches, vaginal rings,
- progestin-only contraceptives – oral pills, intramuscular or subcutaneous injections, subdermal implants,
- intrauterine devices, and
- sterilisation – tubal ligation or insertion of intratubal coils.[9–11]

The profiles of safety and efficacy vary among the different types of contraception. Barrier methods are safe from a cardiac perspective; however, because of the high rates of failure, they are not recommended for women with complex lesions such as hypoplastic left heart syndrome. Women with hypoplastic left heart syndrome are similarly counselled against oestrogen-containing contraceptives because of the associated risk of thromboembolic events with the Fontan circulation. This risk is further increased in women with cyanosis, atrial arrhythmias, or prior thromboembolic events. Progestin-only contraceptives are not associated with a thromboembolic risk and are therefore suitable for women with hypoplastic left heart syndrome. Of the progestin-only systems of delivery, rates of failure of contraception are high with oral pills ("mini-pills"), and these pills are not advised for women in whom pregnancy is considered high risk or is contraindicated. Although intrauterine devices have been used in women with Fontan physiology, the potential exists for vasovagal reaction with insertion leading to reductions in preload and cardiac output. Intrauterine devices should only be placed by gynaecologists with understanding of this condition. Sterilisation may be considered for some women in whom pregnancy is contraindicated; however, this decision is complex. Laparoscopic procedures require insufflation of the abdomen with carbon dioxide, which can potentially be associated with reduction in cardiac output. This reduced cardiac output may be poorly tolerated in women with Fontan circulation. If required, "emergency contraception" is likely safe for women with hypoplastic left heart syndrome.

Counselling about pregnancy

Pregnancy is associated with haemodynamic and hormonal changes that can affect the heart and great vessels and cause an increase in the volume of plasma, an increase in rate of the heart, and an increase in cardiac output. These changes can result in a prolonged haemodynamic load on the heart and subsequent cardiac failure. The systemic right ventricle in women with hypoplastic left heart syndrome may be less able to tolerate the haemodynamic stress of pregnancy and may be prone to cardiac failure when compared with women with a systemic left ventricle. The hormonal changes of pregnancy are thought to contribute to the increased propensity for arrhythmias and changes in the aortic wall. Even when not pregnant, women with Fontan circulation are at increased risk for atrial arrhythmias,[12] and pregnancy further increases this risk. Arrhythmias during pregnancy can be difficult to treat and can be associated with cardiac failure or thromboembolic complications. Furthermore, maternal cardiac failure and arrhythmias are associated with adverse foetal outcomes.[13–15] Pregnancy is also a prothrombotic state, and women with a propensity for thrombosis, such as those with Fontan circulation,[16] are at increased risk for thromboembolic complications. Pulmonary emboli can be particularly dangerous in women with the Fontan circulation, a circulation that requires low resistance for passive flow of pulmonary blood. Residual intracardiac shunts, pulmonary arteriovenous malformations, and systemic-to-pulmonary venous collaterals may result in hypoxaemia, which can have a significant negative impact on foetal growth and development.[17]

In general, the risk of pregnancy in women with Fontan physiology is dependent on several variables:[5–8,13,14]

- functional status,
- degree of cyanosis,
- ventricular function – systolic and diastolic,
- valvar function, and
- the presence of atrial arrhythmias.

Unique risks for women with hypoplastic left heart syndrome relate to the reconstructed aorta and the distorted pulmonary arteries. Reconstruction of the aorta can result in dilation of the neo-aorta. The changes of pregnancy may contribute to progressive dilation or dissection of the neo-aorta as is seen in other aortic pathologies.[18,19] In addition to the risk of maternal cardiac complications during pregnancy, increased risk also exists of adverse maternal obstetric complications, foetal complications, and neonatal complications. Foetal and neonatal complications include premature births, low for gestational age birth weights, and increased incidence of congenital cardiac disease in offspring.[8,13,14,20,21] Owing to these many issues, it is important to educate women that they will

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need comprehensive assessment before conception, and that once they become pregnant they will need care at a centre experienced in the management of congenital cardiac disease in pregnancy. These requirements very often result in the need to consider transfer from a paediatric centre to a centre specialising in adult congenital heart disease.

Transition and transfer

The American Academy of Pediatrics states[1]:

> The goal of transition in health care for young adults with special health care needs is to maximize lifelong functioning and potential through the provision of high quality developmentally appropriate health care services that continue uninterrupted from adolescence through adulthood.

Traditionally, adolescent patients with congenital cardiac disease transfer to a clinic for adults with congenital cardiac disease at 18–21 years of age. The American College of Cardiology task force and the recent "American College of Cardiology/American Heart Association Guidelines on the Management of Adults with Congenital Heart Disease" recommend that the process of transition begins as early as 12 years of age in order to prepare the patient for transition to care as an adult.[22]

For appropriate transition to occur, the patient and family should have a good understanding of the cardiac diagnosis, past interventions, medications, and implications for the future.[23] The patients must understand that they will require life-long follow-up in a clinic for congenital cardiology even if they are feeling well. Patients with repaired hypoplastic left heart syndrome, similar to other patients with functionally univentricular hearts, are a very special group of patients whose management by cardiology needs to occur in the specialised setting of a clinic for adults with congenital cardiac disease. As this cohort continues to age, a broad and mixed skillset will be required to attend to the issues related to congenital cardiac disease, as well as acquired adult cardiac disease.

Counselling

Family planning

It is important that sexual activity, contraception, and reproductive issues be discussed. As previously discussed, a patient with hypoplastic left heart syndrome is considered to be at least at moderate risk for untoward pregnancy, foetal outcomes, and neonatal outcomes.[24]

Genetics

Genetics of congenital cardiac disease continues to be a rapidly evolving area. Approximately 10–15% of newborns with congenital cardiac disease will have an underlying genetic basis for the disease. The majority of the cases are considered to be the result of multi-factorial inheritance.[25] Hypoplastic left heart syndrome is rare, occurring in approximately two of every 10,000 children born. In families who already have one child with hypoplastic left heart syndrome, recurrence rates in siblings was 8%, whereas the risk of a sibling having a hypoplastic left heart syndrome-associated cardiovascular defect was 22%.[26] Resources for genetic evaluations and genetic counselling should be available for assessment and support of these patients and families.

Physical activity

Sports and exercise are an important part of development for all patients and naturally includes those with hypoplastic left heart syndrome.[27] A prescription for exercise should be tailored to the patient based on their diagnosis and functional status. Guidelines from the 36th Bethesda Conference are helpful in determining recommendations for competitive sports and reviewing the classification of sports regarding static and dynamic demands.[28] Diagnostic evaluation before participation in sports can include the following tests:

- a chest radiograph,
- electrocardiogram,
- echocardiography,
- cardiac Magnetic Resonance Imaging, and
- exercise testing with measurement of saturations of oxygen.

Education, employment, and finances

Counselling with regard to education and employment is critical for patients with hypoplastic left heart syndrome. Issues regarding the level of education and physical abilities need to be tailored to the patient. Although illegal, a rational fear of discrimination exists for those with chronic health conditions such as hypoplastic left heart syndrome.[29] Discussion regarding resources in the community and legal resources can be helpful in allaying these concerns.

Health insurance is a problem for many adults with congenital cardiac disease, particularly for the younger adults. In the United States of America, most programmes sponsored by the government are phased out at 18–21 years of age, although recent legislation may delay this problem until the age of 26 years. Up to 30% of adults with congenital cardiac disease who are 19–26 years of age currently have no third-party coverage.[30,31] Support from Social Workers is essential but resources are limited, and obtaining support such as Social Security tends to be a tedious and lengthy process often requiring legal consultation.

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Care coordination

Patients with hypoplastic left heart syndrome should have a provider of primary care who can place an emphasis on adolescent medicine and assist with the process of transition. It should similarly be emphasised that patients see their paediatric cardiologist or their specialist in adults with congenital cardiac disease at least annually, depending on their clinical condition. Surveillance with electrocardiogram, echocardiogram, magnetic resonance imaging, computerised axial tomography, and/or cardiac catheterisation should be utilised as per indications by professionals trained in congenital cardiac disease. Assessment of the cardiac rhythm is carried out by electrocardiogram, Holter, event recording, and electrophysiology study. Long-term issues with ventricular function and arrhythmia, including atrial flutter and/or atrial fibrillation, must be carefully monitored. Furthermore, the potential for re-operation at any stage of the sequence of reconstruction for hypoplastic left heart syndrome must be considered.

Re-operation

Surgical management of patients with hypoplastic left heart syndrome is a three-stage palliative process that starts with the Norwood (Stage 1) operation, usually with a modified Blalock–Taussig aortopulmonary shunt or a Sano right ventricular-to-pulmonary arterial shunt. Stage 1 is usually followed by a bidirectional superior cavopulmonary anastomosis or a hemi-Fontan operation, and then, as a third stage, the completion Fontan operation. The outcome of this three-stage palliation has improved remarkably in the current era because of advances in surgical techniques and neonatal intensive care, with experienced centres reporting excellent overall survival. However, re-operation is inevitable for many patients despite little being reported about outcomes.

Stage I reconstruction: Norwood operation

Coarctation. Recurrent obstruction of the aortic arch after Stage I reconstruction ranges from 9% to 37%,[32] and it can lead to significant ventricular dysfunction and even mortality. Ballweg et al[33] reported a series of 176 patients who underwent Stage 1 reconstruction for hypoplastic left heart syndrome or other variants of functionally univentricular hearts between January 1, 2002, and May 1, 2005 at The Children's Hospital of Philadelphia.

The rate of recurrent obstruction of the aortic arch for all patients in this series was 23% (43 of 176). The rate of recurrent obstruction of the aortic arch for patients alive at more than 30 days of life was 26% (42 of 156). A total of 43 patients underwent balloon angioplasty or surgical intervention. Of the four patients requiring surgical intervention, three had supraaortic obstruction that was intervened upon at the time of the Stage 2 reconstruction. More than one balloon angioplasty was required by seven patients (16%). A total of thirty-nine patients (91%) had balloon angioplasty only between Stage 1 reconstruction and Stage 2 reconstruction, and four patients required balloon angioplasty for recurrent obstruction of the aortic arch both before Stage 2 reconstruction and before Fontan. The median time to surgical or catheter intervention was 123 (1–316) days.

Complications of Right Ventricular-to-Pulmonary Arterial Shunt (Sano Shunt). Ventricular pseudoaneurysm is a rare occurrence after the Sano modification of the Norwood (Stage 1) operation with right ventricular-to-pulmonary arterial conduit. It has been proposed that it is related to the right ventriculotomy that is required for the shunt.[34] Other possible aetiological factors[35] may include:

- pulmonary arterial stenosis at the distal end of the Sano shunt,
- endocarditis,
- technical factors related to incorporating only the epicardial layer in the proximal suture line of Sano, and
- the free pulmonary regurgitation and the systemic right ventricular pressure that may add to the expansion of the aneurysm.

The right ventricular-to-pulmonary arterial shunt provides a more balanced ratio of pulmonary-to-systemic flow of blood and has been reported to improve survival, despite concerns about the long-term outcome regarding the incision in the systemic ventricle and the free pulmonary regurgitation.[36] It has the theoretical advantage of minimising overload of systemic ventricular volume. In a series of 236 infants who underwent Sano modification of the Norwood (Stage 1) operation in the period between June, 2001 and August, 2010, four infants (1.7%) underwent re-operation because of obstruction of the shunt, one died, and three had revisions of the proximal end of the shunt.[37] Although controversial, we agree with the preference of these authors to revise the Sano shunt in these situations – particularly when the re-operation is early – rather than constructing a modified Blalock–Taussig shunt.

Tricuspid valvar regurgitation. One of the aetiological factors behind increased interstage mortality after Stage I reconstruction in infants with hypoplastic left heart syndrome is the development of significant regurgitation of the tricuspid valve, which is the systemic atrioventricular valve. This systemic atrioventricular valvar regurgitation can also lead to failure of the planned staged palliative reconstruction. Tricuspid regurgitation can result from right

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ventricular dysfunction or an intrinsic abnormality of the tricuspid valve, or it may result from a combination of both of these aetiologies. Valvar repair provides an option to preserve right ventricular function and improve survival. Repair of the tricuspid valve can be performed as a separate procedure or as a part of the second or the third stage procedures. Bove et al[38] presented the results of repair of the tricuspid valve in 28 patients who underwent staged reconstruction for hypoplastic left heart syndrome in the period between 1994 and 2002.[38] These 28 patients represented 6% of all their patients with hypoplastic left heart syndrome who underwent reconstructive surgery during that period of time. In this series, several techniques were used to repair the tricuspid valve:

- partial eccentric annuloplasty creating bileaflet closure was the most frequently used technique,
- in six patients with localised areas of prolapse, the authors applied the technique of partial closure of zones of apposition with adjacent leaflets, and
- in five patients, ringed annuloplasty was performed.

On late follow-up, 17 patients had a successful repair with mild or less tricuspid regurgitation, and 15 underwent completion Fontan procedure. Importantly, of those who did not have a successful repair of the tricuspid valve, eight patients died. Overall survival was 67%, and in those who underwent successful repair of the tricuspid valve, the survival rate was 94%.

Stage II reconstruction: bidirectional superior cavopulmonary anastomosis

Occlusion of the cavopulmonary anastomosis. Occlusion of the cavopulmonary anastomosis represents a catastrophic complication that warrants a high index of suspicion, which will lead to an early diagnosis and aggressive intervention to prevent mortality. In four infants ranging from 4 to 8 months of age who presented after their second stage reconstruction with acute occlusion of the cavopulmonary connections,[39] all required emergency catheterisation that confirmed the occlusion. The time of thrombosis was between 3 and 5 days post-operatively. The left pulmonary artery and the left-sided superior caval vein were the main sites of occlusion. Although re-operation is an option, these four infants were successfully managed with stenting of the occluded segment. Owing to recurrent occlusion of the left pulmonary artery, one patient died. Risk factors as outlined by the authors included:

- a state of low cardiac output,
- younger age at surgery with smaller size vessels,
- indwelling central venous catheters, and
- stasis secondary to residual obstruction.

Stage III reconstruction: Fontan operation

Fontan takedown. Iyengar et al[40] reported three patients with hypoplastic left heart syndrome who underwent takedown of their Fontan in the period between 1980 and 2007. These three patients underwent their completion extracardiac Fontan at 3, 6, and 15 years of age. Their main reasons behind failure of the Fontan were

- the development of systemic-to-pulmonary collaterals,
- pulmonary venous obstruction, and
- diastolic dysfunction, which led to mortality in one patient.

All three patients presented with pleural effusion, and the Fontan was taken down to a bidirectional cavopulmonary anastomosis. It is difficult to reach conclusions from this small series, but the authors recommended takedown of the Fontan in patients with persistent effusions within 6 weeks to avoid deterioration of their nutritional status.

Fontan conversion and arrhythmia surgery. "Fontan conversion" is one way of managing patients with failing Fontan circulation. The principles of Fontan conversion include:

- conversion of prior atriopulmonary Fontan to total extracardiac cavopulmonary connection,
- surgical treatment of underlying arrhythmias,
- surgical management of any associated systemic valvar dysfunction, which is usually tricuspid regurgitation, and
- treatment of all obstructions of the systemic or pulmonary venous pathways.

Right atrial re-entry tachycardia is managed by a right-sided Maze procedure, whereas atrial fibrillation and left-sided atrial re-entry tachycardia are managed by a left-sided Maze procedure.

Neo-aortic root aneurysm after Fontan completion. Augmentation of the native aorta with a patch is usually an integral part of the aortic reconstruction in the Norwood (Stage 1) operation. The long-term durability of such patches is unknown, despite some experimental data that reported the development of aneurysms with the use of pulmonary homografts.[41] Ehsan et al[42] reported a 14-year-old boy who underwent a Norwood (Stage 1) operation at 3 days of life with a pulmonary homograft, and at 17 months of age he completed his reconstructive stages with a lateral tunnel fenestrated Fontan. During follow-up, his neo-aorta progressively increased in

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size, and he underwent re-operation with replacement of his aneurysmal aorta, with performance of the distal anastomosis under deep hypothermic circulatory arrest. The histological examination of the aorta revealed evidence of cystic medial degeneration, whereas the pulmonary homograft showed only a thin rim of elastic fibres with dense fibrous scarring in the adventitia. A similar situation has been observed in patients after the Ross operation with exposure of the pulmonary valve and pulmonary artery to systemic arterial pressure;[43] after the Ross operation, the potential for aneurysmal dilation of the neo-aortic root and neo-aortic valvar regurgitation exists.

Valve-sparing replacement of the neo-aortic root represents an option for these children with dilation of the neo-aortic root. Pizarro et al[44] reported a valve-sparing replacement of the neo-aortic root in a 10-year-old child after completion Fontan procedure for hypoplastic left heart syndrome. The child had an aneurysm that measured 5.4 centimetres at the level of the sinuses of Valsalva, with a competent aortic valve. The aneurysm caused near-complete obliteration of the left pulmonary artery. Fragmentation with loss of elastic fibres, and smooth muscle cells with deposition of myxoid material in the media, were the main findings on histopathological evaluation. The material of the patch used in the aortic reconstruction during the initial palliative stage may have an effect on future development of an aneurysm of the neo-aorta; however, with the development of the aneurysm proximal to the homograft reconstruction, a different mechanism may be behind the formation of the aneurysm. Histologically, the true native pulmonary root is not identical to that of the true native aortic root. Thus, structurally, the true native pulmonary root may be prone to dilation under life-long systemic pressure and impedance. This same situation exists for the Ross autograft.

Transplantation

Cardiac transplantation is still an option for patients with hypoplastic left heart syndrome, even with the marked improvement in the surgical and medical strategies of management involving staged reconstruction.

Aortic complications. Overall, aortic complications are uncommon after cardiac transplantation. The reported incidences are 0.35%[45] and 2%[46] in two series. During the Norwood (Stage 1) operation, the hypoplastic ascending aorta and the aortic arch are augmented with a patch of homograft. Little data exist about the fate of these patches. Kanter et al[47] presented two patients out of 16 children who underwent orthotopic cardiac transplantation after previous Norwood. The first patient underwent re-operation for an aortic pseudoaneurysm that caused secondary airway compression. The second one had recurrent coarctation after transplantation and underwent multiple balloon dilations. After 18 years, the patient developed a contained mycotic pseudoaneurysm of the reconstructed transverse aortic arch that required replacement of the reconstructed aorta.

Prevention of post-operative pericardial adhesions

With improved outcome and survival in children with hypoplastic left heart syndrome, re-operation is becoming an expected part of the surgical care of these patients. Post-operative adhesions increase the risk with re-operation. Use of membranes made of polytetrafluoroethylene as substitutes for the pericardium is one of the strategies to facilitate sternal re-entry.[48] Salminen et al[49] presented a prospective randomised comparative study that included 21 patients with hypoplastic left heart syndrome. Membranes made of polytetrafluoroethylene and synthetic barriers to adhesions using polyethylene glycol hydrogel were applied just before sternal closure during stage 1 palliation. The authors reported that the application of these membranes and synthetic barriers to adhesions is safe and technically easy. The membrane made of polytetrafluoroethylene alone or with the synthetic barrier to adhesions using polyethylene glycol prevented adhesions between the heart and the sternum; however, the density of adhesions tended to increase. No significant reduction in the formation of pericardial adhesions resulted with the use of synthetic barriers to adhesions using polyethylene glycol alone. The authors concluded that a slight benefit might exist in using the membranes made of polytetrafluoroethylene.

Conclusion

In conclusion, caring for patients with hypoplastic left heart syndrome can be challenging and rewarding. Thankfully, over the last quarter of a century or so, this group of patients has gone from 0% survival to about 75% survival. New and unforeseen issues are likely to arise as more patients with hypoplastic left heart syndrome enter into young adulthood, and our oldest cohort moves into middle adulthood. As a group of paediatric cardiologists, cardiovascular surgeons, cardiac anaesthesiologists, cardiac intensivists, cardiac nurses, perinatologists, neonatologists, and specialists in adults with congenital cardiac disease specialists, we will continue to strive for better outcomes with improved quality of life for this unique and pioneering population of patients.

The manuscript considers multiple factors of concern for long-term survivorship of patients with hypoplastic left heart syndrome. This manuscript does not, however, cover the long-term medical

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complications of the Fontan circulation, which may indeed dominate all the other concerns. These long-term medical complications after the Fontan operation are discussed in detail elsewhere in this HeartWeek 2011 Supplement of Cardiology in the Young in the following two manuscripts:

- *New concepts*: Development of a Survivorship Programme for Patients with a Functionally Univentricular Heart by David J. Goldberg, MD et al, and
- *Long-term management of patients with hypoplastic left heart syndrome*: The Diagnostic Approach at All Children's Hospital by Gul H. Dadlani, MD et al.

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