**Cardiovascular CT for Evaluation of Single Ventricle Heart Disease: Risks and Accuracy Compared to Interventional Findings**

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Objective:

We sought to evaluate risk and image quality of cardiovascular computed tomography (CT) in patients through all stages of single ventricle palliation, and to define accuracy by comparing findings to intervention and surgery.

Methods:

Consecutive CT scans performed in patients with single ventricle heart disease were retrospectively reviewed at a single institution. Diagnosis, sedation needs, estimated radiation dose and adverse events were recorded. Anatomic findings, image quality (1-4, 1=optimal), and discrepancy compared to interventional findings were determined. Results are described as median (25th, 75th percentiles).

Results

From January 2010 - August 2015, 132 CT scans were performed in single ventricle patients (20 neonates, 52 post Norwood, 15 post Glenn and 45 post Fontan). No sedation was used in 76 patients, 47 were under minimal or moderate sedation, and 9 were under general anesthesia. Median image quality score was 1.2. Procedural dose length product was 24 milligray-centimetersand unadjusted/adjusted radiation doses were 0.34 (0.2, 1.8) and 0.82 (0.55, 1.88) millisievert. There was one adverse event. No major and 2 minor discrepancies were noted at the time of 79 surgical and 10 catheter based interventions.

Conclusions:

Cardiovascular CT can be performed with a low radiation exposure for patients with single ventricle heart disease. Accuracy compared to interventional findings is excellent. CT is an effective advanced imaging modality when a noninvasive pathway is desired, particularly if cardiac MRI is high risk or contraindicated.

**Introduction**

A majority of patients with single ventricle heart disease will survive to adulthood1-3. These patients need anatomic evaluation between palliative procedures and intermittently throughout their lives. Echocardiography is insufficient for evaluation of thoracic vasculature or for reproducible estimation of ventricular function4-6. Cardiovascular magnetic resonance imaging (MRI) is commonly performed for this indication but requires relatively long imaging times, deep sedation or anesthesia for young children. Many older patients have metallic implants with artifact that degrade MRI image quality.7 In addition, it is relatively contraindicated in those with the most current generation pacemakers and defibrillators and the devices have been known to cause imaging artifact8. Recent findings of cerebral gadolinium deposits recommend MRI use be carefully considered 9-11. Cardiovascular computed tomography (CT) has been shown to be accurate for evaluation of anatomy and function for most indications in congenital heart disease12, but there has not been a report of image quality or a correlation to interventional findings in a cohort of single ventricle patients through all stages of palliation.

**Methods**

A review of cardiovascular CT scans performed in patients with single ventricle heart disease from a prospective imaging database at a single institution was performed. Institutional review board approval was obtained.

Patient demographics

Patient age, underlying cardiac diagnosis, height and weight, non-sedated vs sedation vs intubation, IV site and gauge, and adverse procedural events were determined from patient records. Patients were grouped by stage of palliation into four groups including neonate prior to intervention and status post Norwood, Glenn and Fontan.

Cardiac computed tomography indication

The indication for cardiovascular CT rather than cardiac MRI was grouped as follows: 1) patients in anesthesia class 3-4 considered relatively high risk for anesthesia and CT could be performed with limited or no anesthesia, 2) poor MRI image quality due to metallic artifact or presence of defibrillator or pacemaker, 3) need for extracardiac and/or high resolution coronary artery imaging, or 4) claustrophobia or obesity and unable to fit in MRI scanner.

Scanner platform and scan sequence, patient preparation

Studies were performed on a second generation dual source CT scanner (Somatom Definition Flash, Siemens Healthcare, Forchheim, Germany, gantry rotation time = 280 milliseconds, temporal resolution = 75 milliseconds, collimation of 2 x 128 x 0.6 millimeter). For anatomic imaging, a prospectively electrocardiogram triggered high pitch (3.4) helical scan was performed using automated online tube current modulation. For the scanner platform used, electrocardiogram gating is needed to get the highest pitch scan mode, therefore leads were placed or the “demo mode” of electrocardiogram gating was used on the scanner to allow use of highest pitch without placement of electrocardiogram leads on the patient. For patients older than one year, electrocardiogram leads were typically placed. For functional imaging a retrospectively electrocardiogram -gated helical scan sequence (spiral) was performed using a minimal diastolic acquisition window, automated online tube current modulation and electrocardiogram pulsed modulation. For all patients the tube potential was adjusted lower if the automated software picked a higher kilovolt peak than needed based on clinical judgement. In 2011, 70 kilovolt peak tube potential became available with a scanner upgrade. Scans were reconstructed using the Siemens second generation iterative reconstruction algorithm, Safire, at a strength of 3.

All neonates and infants (pre-Norwood, status post stage 1 and 2) underwent an anatomic scan in prospectively electrocardiogram triggered high pitch scan mode with a single scan or two scans performed back to back. Early in the experience a single scan was performed. For neonates, pre-Glenn and post-Glenn patients, contrast dose of 2 ml/kg was injected at the rate appropriate for age and intravenous gauge. Due to the variability of both contrast timing and intracardiac shunting, there were times that dense scatter artifact on a single scan made it difficult to differentiate artifact from pathology, or a scan was performed that did not optimally opacify both systemic venous and arterial anatomy simultaneously and central venous occlusion was unable to be evaluated. To improve definitive assessment of both systemic and pulmonary arterial and venous anatomy, two scans were performed in most neonates and infants prior to the Glenn cavopulmonary anastomosis in the recent era.

Power injection of contrast was done through a 24 - 20 gauge catheter based on patient size in most cases, with few patients having injection through a broviac or central venous catheter when peripheral intravenous access was unable to be obtained. Intravenous site was typically chosen by the practitioner placing the line, occasionally requested in a certain position if possible (example: lower extremity intravenous access for a patient with interrupted inferior vena cava). 22 gauge intravenous catheter or larger was requested in all patients, and when not able to be achieved, a 24 gauge intravenous catheter was used in neonates and infants. Power injection was used for contrast administration after a hand injection of saline and a test bolus of saline at the anticipated contrast injection rate or slightly higher to account for the increased viscosity of contrast compared to saline. Rarely, if the intravenous line was considered fragile or positional, a hand injection was used. Scans were initiated based on visualization of contrast in the structure of interest on a monitoring sequence, which was initiated approximately halfway through the contrast injection. Scan range included the thorax, extended cranially for those requiring airway evaluation, and caudally to include the upper abdomen in those with possible aortopulmonary collateral vessels. Most patients following a Fontan underwent an electrocardiogram triggered functional computed tomography scan with electrocardiogram based tube current modulation and a narrow diastolic acquisition window, followed 30-60 seconds later by an anatomic scan using a prospectively electrocardiogram triggered high pitch helical scan protocol. The functional scan was performed to allow quantification of single ventricle function or to visualize contrast and unopacified venous streaming and admixture in patients after the Fontan procedure, which is often confused for clot in a single phase image. For Fontan patients, the contrast was given in two phases with a 30-60 second pause between contrast injections, with a longer pause for those with significant valve regurgitation or decreased ventricular function. The monitoring sequence was timed to the second phase of the contrast bolus, and the functional image acquisition was timed to opacification of the cardiac structures on the monitoring sequence. The anatomic scan was performed 40-60 seconds later to optimize the opacification of the Fontan pathway. A longer pause between scans was used for those with significant valve regurgitation, decreased ventricular function or atriopulmonary Fontan where cardiac output and contrast timing were predicted to be later than usual. A pediatric (KH) or adult cardiologist (JL) experienced in congenital CT was present for all scans. A neonatal nurse practitioner team transported all neonates coming to the scanner from the neonatal intensive care unit and were present during the scan. An anesthesia team transported and was present for intubated patients of all ages coming from an intensive care unit, and for intravenous line placement and sedation when needed for all patients less than 6-8 years of age. Outpatients over 8 years of age presented directly for scanning to the hospital based imaging center.

Sedation and anesthesia use

Neonates were scanned without sedation or anesthesia unless required for clinical management of underlying congenital heart disease. Patients after first stage palliation were managed by the anesthesia team and were scanned without sedation whenever possible. Patients were scanned free breathing with minimal sedation when required for cooperation or intravenous line placement. Sedation goal was for the patient to be comfortable, alert and interactive during the exam, and bundled to restrict patient motion. Critically ill patients were intubated for management of their underlying congenital heart disease (CHD). Children less than 6 years old who required functional data or high resolution coronary imaging were placed under general anesthesia with intubation for breath holding during image acquisition, since sequences for both indications acquire data over several heart beats and respiratory motion compromises image quality.

Image Quality

Images were reviewed qualitatively on a four point scale (1= excellent image quality with optimal visualization of all anatomic targets, 2= good image quality with diagnostic visualization of all anatomic targets, 3= marginal image quality with diagnostic visualization of most anatomic targets, 4= poor image quality, nondiagnostic for evaluation of anatomic targets). Image quality included assessment of diagnostic visualization of all anatomic targets for the entire exam based on the underlying anatomy and stage of palliation. For all scans with a score > 1, the reason for the suboptimal image quality was determined such as poor contrast or motion artifact. Quantitative image quality was determined by calculation of noise, signal to noise ratio and contrast to noise ratio in the systemic artery.

Radiation dose parameters:

The scanner platform, contrast, imaging sequence, CT dose volume index (milligray), scan dose length product (milligray-centimeters, 32 cm phantom), scan length, tube potential and tube current were recorded for each scan. Individual scan and cumulative procedural dose length product in milligray-centimeters were recorded.

Radiation Dose Estimation

Procedural dose length product was used to estimate radiation dose. An unadjusted radiation dose in millisievert was calculated by multiplying dose length product by the standard chest conversion factor (scan dose length product x .014)13. For patients <18 years of age, conversion factors were additionally calculated by age (0.039 for ≤0.50 years, 0.026 for 0.51 to 2.50 years, 0.018 for 2.51 to 7.50 years, and 0.014 for patients >7.50 years) 14-15.

Adverse events and Correlation to Interventional Findings

Patient records were reviewed for adverse event with scanning or recovery. Subsequent catheterization and surgical reports were reviewed for discrepancy between the pre and post procedural diagnosis. Inpatient and outpatient records through subsequent intervention were additionally reviewed for diagnostic discrepancy.

Statistical Methods

Descriptive statistics are displayed as median (25th, 75th percentile) for continuous variables unless otherwise stated; number and percentage with characteristic are reported for categorical variables. Continuous variables were analyzed using Kruskal-Wallis tests and categorical variables were analyzed using Pearson’s chi-square or Fisher’s exact tests. A value of p<0.05 was considered significant, and p-values are two-sided where possible. P-values for pairwise comparisons were adjusted using the Bonferroni method. All statistical calculations and plots were done with Stata 14.1 (StataCorp LP, College Station, TX).

**Results**

Indication for computed tomography by patient group

During the time of review, 132 cardiovascular CT scans were performed in patients with single ventricle heart disease. Underlying cardiac diagnoses for all patients was hypoplastic left heart syndrome in 44%, right sided heart disease including tricuspid atresia, pulmonary atresia and Ebsteins anomaly in 26% of patients, complex heterotaxy/atrial isomerism in 20%, and double inlet ventricle in 10%.

Twenty neonates underwent evaluation for the following indications: pulmonary venous anomaly (n=14), branch pulmonary artery anomaly (n=5) or aortic arch for possible interruption (n=1). CT was performed in neonates rather than other diagnostic modality since they are considered relatively high risk for anesthesia and CT could be performed with limited or no sedation/anesthesia.

Fifty-two patients were referred for CT imaging between stage 1 and 2 single ventricle palliation. In this group, 44 patients were referred for standard pre-Glenn imaging because they were considered low risk for requiring catheter intervention based on echocardiogram and clinical assessment. For this group of patients the CT scan replaced the prior practice standard of cardiac catheterization prior to second stage palliation. Alternatively, pre-Glenn patients were referred for CT imaging prior to the time of routine pre-Glenn evaluation if there were concerns regarding anatomy (arch, venous collaterals, pulmonary artery stenosis) based on either clinical status or echocardiogram (n=8). For all patients electively imaged between stage 1 and 2 palliation, CT imaging was pursued rather than MRI because it could be performed with limited sedation, this group is relatively high risk for adverse events with anesthesia, and the average MRI imaging time is relatively long. For patients urgently imaged, the length of anesthesia was taken into consideration in determining the diagnostic modality.

Fifteen studies were performed after second stage cavopulmonary anastomosis and before Fontan palliation. Patients were referred for concerns regarding anatomy based on either clinical status or echocardiogram. In this group, 4 patients had pacemakers and in the remainder CT imaging was pursued rather than MRI because it could be performed with limited or no anesthesia. The practice standard during this time remained invasive diagnostic catheterization prior to third stage Fontan palliation.

Forty-five patients were referred for anatomic evaluation after Fontan completion as part of lifelong interval assessment. The practice standard for post-Fontan evaluation during this time remained non-invasive evaluation, primarily utilizing cardiac MRI. Of the patients who underwent CT, 42 had pacemaker or prior MRI with artifact, 2 required detailed coronary artery imaging, and one required evaluation of a chest mass in addition to cardiac anatomy.

Use of sedation and intubation:

For the total group, 76 patients were scanned without sedation, 47 were scanned with minimal to moderate sedation, and 9 patients were intubated during the scan. Of the 9 patients intubated during the scan, 6 were intubated for clinical reasons, and 3 patients were intubated specifically for the scan (2 for multiple procedures, and one for the cardiac CT alone).

Eighteen patients in the neonatal group were scanned without sedation and two were intubated for clinical management of underlying heart disease. Of the 52 patients scanned between first and second stage palliation, 13 patients were scanned without sedation, 34 were scanned free breathing but with sedation, and 5 were intubated. Three of these five patients were intubated for clinical reasons and two patients were intubated with general anesthesia for multiple concurrent procedures (head CT, cardiac CT on different scanners in a patient with cyanosis; cardiac CT and gastric tube placement in the other patient). Of the 15 patients scanned between stage 2 and 3 palliation, two were not sedated, 12 were sedated but free breathing, and 1 was intubated for clinical reasons. In the 45 patients undergoing CT scan after Fontan palliation, 43 were scanned without sedation, one patient with developmental delay was sedated for the scan and one patient with developmental delay and movement disorder was intubated for a breath hold sequence.

Intravenous Line site and Gauge:

Information on intravenous line site and gauge was available in 128 of the 132 scans (97%). Three patients had hand injection of contrast through a central line, 1 patient had power injection through a broviac catheter, 1 neonate had hand injection through an umbilical venous catheter, and 1 patient had hand injection through a Peripherally Inserted Central Catheter line when other access unable to be obtained. The remaining 122 patients had contrast injection through a peripheral line. Table 1 lists the peripheral intravenous gauge by stage of palliation for these patients. Peripheral intravenous line location was a right or left sided hand vein, right or left sided antecubital vein, or a right or left sided saphenous vein.

Scan Sequences Used by Patient group

In the neonatal group (n=20) 6 patients underwent a single anatomic scan and 14 underwent two anatomic scans, for a total of 34 anatomic scans. No patient underwent functional imaging. Twenty-seven anatomic scans were performed using tube potential of 70, and 7 anatomic scans were performed using tube potential of 80.

For the patients scanned between first and second stage palliation, 28 had a single anatomic scan and 24 underwent two anatomic scans, for a total of 76 scans. No patient underwent functional imaging. Thirty-nine scans were performed using tube potential of 70, and 37 scans were performed using tube potential of 80.

For the patients scanned between the Glenn and Fontan (n=15), 6 patients had a single anatomic scan, 6 had two anatomic scans, and 3 had an anatomic and a functional scan, for a total of 21 anatomic scans and 3 anatomic scans. 13 anatomic scans were performed using tube potential of 70, and 8 anatomic scans were performed using tube potential of 80. One functional scan was performed using tube potential of 70, and two functional scans were performed using tube potential of 80. After the Fontan procedure (n=45) two patients underwent a single anatomic scan, one patient had a single functional scan, 4 patients had two anatomic scans, and 38 patients had an anatomic and a functional scan performed, for a total of 48 anatomic scans and 39 functional scans. 18 anatomic scans were performed using tube potential of 70, and 27 anatomic scans were performed using tube potential of 80, and three anatomic scans were performed using a tube potential of 100. 4 functional scans were performed using tube potential of 70, 22 functional scans were performed using tube voltage of 80, 12 functional scans were performed using a tube potential of 100 and 1 scan was performed using a tube potential of 120.

Estimated radiation dose

Table 2 lists procedural dose length product and radiation dose estimates for the entire group stratified by stage of palliation. Median procedural dose length product was 24 milligray-centimeters (16, 114) and unadjusted and adjusted radiation doses were 0.34 (0.20, 1.80) and 0.82 (0.55, 1.88) millisievert, respectively.

Image Quality

Qualitative image quality for the group showed an image score of 1 for 118 of the 132 scans (89%), an image score of 2 for 12 scans (9%) and an image score of 3 for 2 scans (2%). The 2 scans which received an image score of 3 were both in the post Fontan group. Of the 14 scans with image quality score less than 1, 8 had poor contrast opacification in the structures of interest due to scan timing, 2 had intravenous contrast leak with poor contrast in the structures of interest, 2 had contrast scatter artifact, and 2 had movement artifact. Table 3 summarizes both qualitative and quantitative image quality data.

Adverse events, significant anatomic findings and correlation to intervention

There was one adverse event of a broviac catheter (placed two months before) crack with contrast injection that was able to be repaired. Multiple attempts at peripheral intravenous placement were unsuccessful in this case. A power injector was used for contrast injection at a rate of 1 milliliter per second and the pounds per square inch was acceptable with saline test injection but over the recommended pounds per square inch for the catheter with a 50%/50% contrast/saline mix at the same injection rate. The catheter crack was repaired and the catheter remained functional through hospital discharge.

In total, 79 patients underwent surgical intervention after CT scan, 7 patients underwent catheter based intervention prior to surgery, and 3 patients underwent catheterization based intervention alone.

Of the 20 neonates, 18 underwent subsequent staged palliation and two patients died. No patient had additional advanced diagnostic studies prior to initial surgical palliation. A minor discrepancy was a difference of opinion regarding the classification of a vessel as an aortopulmonary collateral vs ductus arteriosus. Example neonatal images are shown in image 1.

Of the 52 patients who were scanned after first stage single ventricle palliation, significant findings other than expected anatomy were 7 central venous occlusions, 2 aortopulmonary shunt or Sano narrowings > 50%, 12 significant branch pulmonary artery narrowing, 2 arch obstruction, and 1 large pseudoaneurysm at the proximal Sano insertion. There were 50 patients who underwent subsequent surgical intervention and two patients who died; one during anesthesia induction for planned catheter based arch angioplasty and one with progressive and severe pulmonary vein stenosis. One surgical intervention was a hybrid procedure with stent placement at the time of Glenn. Four patients underwent catheter based intervention prior to surgery including pulmonary artery balloon angioplasty, pulmonary artery balloon angioplasty with stent placement (n=2), and aortic arch balloon angioplasty. There was one discrepancy compared to interventional findings which was the presence of right internal jugular vein occlusion in a patient who underwent catheterization two months subsequently. The CT scan in that patient was not optimized for venous imaging and only a single scan was performed. Examples of pre-Glenn images are shown in image 2.

Of the 15 patients imaged between the Glenn and pre-Fontan catheterization, 3 instances of central venous occlusion, one pulmonary artery narrowing, 2 arch obstruction, and two large venous collaterals were found. Five patients underwent intervention (3 catheterization, 2 surgical) prior to third stage palliation. Interventions included pulmonary arterioplasty, aortic arch angioplasty, aortic arch augmentation, and coiling of large venous collaterals (n=2). There were no discrepancies in this patient subset compared to interventional findings. Examples of images after the Glenn procedure are shown in image 3.

Anatomic findings in 45 post Fontan patients included 6 complete central venous occlusion with large decompressing collaterals, 1 arch obstruction, bulboventricular foramen restriction, an anomalous left coronary artery with interarterial course, and a near occlusive clot in the superior vena cava. Twelve patients underwent intervention (9 surgical, 3 catheter based). Surgical interventions included unroofing of a coronary artery and pulmonary valvuloplasty, bulboventricular foramen enlargement, tricuspid valvuloplasty, neo-aortic valve replacement, conversion to extracardiac Fontan, and 4 pacemaker generator changes. Catheter based interventions included clot removal, arch angioplasty and stent with vascular plug of collaterals, and vascular plug and coil occlusion of collaterals. There were no discrepancies in this subset of patients compared to interventional findings. Example images after the Fontan procedure are shown in image 4.

**Discussion**

CT is a robust alternate diagnostic modality through all stages of single ventricle palliation. For highly select indications, the risk profile may be preferable to other modalities if radiation, vascular access and anesthesia risks are considered. Single ventricle patients have a relatively high cumulative radiation exposure through staged palliation16,17. A single institution survey reports a median cumulative effective radiation dose of 25.7 millisievert from birth to 33 months of age, 78% from catheterization which constituted 4% of radiation encounters16 . Another survey of cumulative radiation dose for patients with all forms of congenital heart disease showed 5.3% of patients received over 20 millisievert/year with a median follow-up of 4.3 years18. A recent study directly comparing radiation dose from diagnostic catheterization (n=50 cases) and CTA (n=50 cases) in pediatric patients with CHD has shown 15 fold less radiation for computed tomography angiography, although not specific to patients with single ventricle heart disease19. Other studies using older CT equipment list doses for computed tomography angiography (n=21) two fold higher than diagnostic cardiac catheterization (n=117)17. These results show the radiation risk of CT varies considerably depending on the scanner platform used and the aggressiveness of dose reduction. The image quality necessary for evaluation of coronary lesions in adult patients is rarely needed for congenital applications and patient specific dose reduction must be implemented if a diagnostic strategy utilizing CT implemented. The standard diagnostic protocol at a majority of centers remains invasive catheterization prior to both second and third stage palliation despite data showing favorable risk profile for non-invasive evaluation3.

Non-invasive assessment prior to second stage palliation has shown similar operative outcomes compared to invasive catheterization with lower risk as measured by radiation exposure, vascular access complications, length of anesthesia and adverse events20-22. Longer term follow-up shows no difference in outcomes between invasive and non-invasive evaluation prior to stage 2, measured up to eight years after the Fontan procedure. Our practice now uses CT preferentially for evaluation of anatomy prior to second stage palliation. Catheterization is reserved for patients for whom intervention is considered likely based on echocardiogram or clinical exam, and for patients with poor ventricular function and severe valve regurgitation for whom hemodynamics are considered relevant to clinical management. Some experts now propose a noninvasive algorithm for evaluation prior to both second and third stage palliation in patients with single ventricle heart disease considered low risk for requiring intervention23-25.

Cardiovascular MRI is the most commonly used noninvasive advanced imaging modality in CHD, but deep sedation or general anesthesia is required for young children, scan times are relatively long, and gadolinium is used in many patients for angiography. Anesthesia has increased risk in patients with complex congenital heart disease undergoing MRI evaluation and there is concern that repeated anesthesia in young patients may have adverse neurological effects26-35. Gadolinium deposits have been found in brain tissue after repeat dosing in both pediatric and adult patients, the significance of which is not yet known36-39. Risk assessment of noninvasive modalities should include assessment of anesthesia risk, iodinated or gadolinium based contrast exposure in addition to radiation exposure and vascular access requirements.

Cardiovascular CT is an alternate imaging modality that can be used as part of the noninvasive pathway when cardiac MRI is considered high risk or there is imaging artifact40. When anesthesia is needed, a single breath hold is required for data acquisition and the length of anesthesia will be relatively short. Function analysis for both right and left ventricles has been shown to be comparable to cardiac MRI when using CT scanners with acceptable temporal and spatial resolution41-45.

**Limitations**

Findings from this study are retrospective and limited to a single institution. Accuracy compared to interventional findings is limited to structures present within the surgical field or imaged by invasive angiography. Additionally, these findings may not be generalized to institutions with different CT professional and hardware capabilities.

**Conclusion**

Cardiovascular CT offers an alternate noninvasive imaging modality for patients with single ventricle heart disease when advanced imaging is preferred but for whom cardiac MRI is high risk, has poor image quality, or is contraindicated. Image quality remains good at a low radiation exposure and accuracy compared to interventional findings is excellent. CT may reduce radiation risk compared to catheterization and decreases anesthesia and gadolinium risk compared to MRI. It should have a role in select patients with single ventricle heart disease when appropriate staff and CT equipment are available.

Figure Legend

Figure 1 – Neonatal Evaluation

1A: 2D image of total anomalous pulmonary venous return with a vertical vein (arrow) to the innominate vein in a patient with hypoplastic left heart syndrome.

1B: Anomalous right pulmonary vein (arrow) and an aortopulmonary collateral (short arrow) in a patient with Scimitar syndrome and hypoplastic left heart syndrome.

1C: 3D image of bilateral superior vena cava (arrows) in a patient with tricuspid atresia.

Figure 2 – Post Norwood evaluation

2A: 3D reconstruction of an aortopulmonary shunt (arrow) from the base of the right innominate artery to the right pulmonary artery. Left SVC is noted.

2B: Axial 2D image of a distal Sano anastomosis to the branch pulmonary arteries.

2C: Saggital 2D image of an occluded aortopulmonary shunt (\*\*\*) from the base of the innominate artery (arrow).

2D: Posterior view of a 3D reconstruction showing aortic coarctation at the distal Norwood anastomosis (arrow) after first stage Sano palliation.

Figure 3 – Post Glenn Evaluation

3A: Coronal 2D image of narrowing (arrow) of the superior vena cava to right pulmonary artery anastomosis after cavopulmonary connection.

3B: Coronal 2D image showing a clot in the superior vena cava (arrow) and a patent left pulmonary artery stent (\*) after Glenn cavopulmonary anastamosis.

3C: Coronal 2D image showing dense contrast opacification of decompressing venous collaterals (arrow) in a patient with innominate vein and subclavian vein occlusion after glenn cavopulmonartery anastomosis..

3D: Saggital 2D reconstruction in the same patient showing a patent LPA stent (arrow) on a venous phase acquisition.

3E: Long segment tracheal atresia (\*\*\*) in a patient after the Glenn procedure. Both the proximal and distal tracheal are visualized (arrows).

Figure 4: Post Fontan Evaluation

4A: Coronal 2D image from the venous phase scan in a patient after lateral tunnel Fontan. The Fontan fenestration patch is noted (\*).

4B: 2D image of retained pulmonary valve leaflets (arrow) in a patient after pulmonary artery ligation. The pathway from the systemic LV to the aorta is mildly narrowed (\*).

4C: 2D axial image of apatent Fontan fenestration stent (\*) with unopacified flow from the Fontan to the atrium (arrow) after septectomy.

4D: 3D reconstruction of a densly opacified large decompressing collateral from the innominate vein after the Fontan procedure (\*\*\*).

4E: 2D coronanal image of both veno-venous (arrows) and arterial collaterals(\*) after the Fontan procedure.

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1. Khairy P, Fernandes SM, Mayer JE, Jr., Triedman JK, Walsh EP, Lock JE, et al. Long-term survival, modes of death, and predictors of mortality in patients with Fontan surgery. Circulation. 2008;117(1):85-92.

2. Khairy P, Ionescu-Ittu R, Mackie AS, Abrahamowicz M, Pilote L, Marelli AJ. Changing mortality in congenital heart disease. Journal of the American College of Cardiology. 2010;56(14):1149-57.

3. Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease). Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. Journal of the American College of Cardiology. 2008;52(23):e143-263.

4. Banka P, McElhinney DB, Bacha EA, Mayer JE, Jr., Gauvreau K, Geva T, et al. What is the clinical utility of routine cardiac catheterization before a Fontan operation? Pediatric Cardiology. 2010;31(7):977-85.

5. Weiss F, Habermann CR, Lilje C, Sasse K, Kuhne T, Weil J, et al. [MRI in postoperative assessment of univentricular heart disease: Correlation with echocardiography and angiography]. RoFo : Fortschritte auf dem Gebiete der Rontgenstrahlen und der Nuklearmedizin. 2002;174(12):1537-43.

6. Stern KW, McElhinney DB, Gauvreau K, Geva T, Brown DW. Echocardiographic evaluation before bidirectional Glenn operation in functional single-ventricle heart disease: comparison to catheter angiography. Circulation Cardiovascular Imaging. 2011;4(5):498-505.

7. Garg R, Powell AJ, Sena L, Marshall AC, Geva T. Effects of metallic implants on magnetic resonance imaging evaluation of Fontan palliation. The American Journal of Cardiology. 2005;95(5):688-91.

8. Cronin EM, Mahon N, Wilkoff BL. MRI in patients with cardiac implantable electronic devices. Expert review of medical devices. 2012;9(2):139-46.

9. Ramalho J, Semelka RC, Ramalho M, Nunes RH, AlObaidy M, Castillo M. Gadolinium-Based Contrast Agent Accumulation and Toxicity: An Update. AJNR American Journal of Neuroradiology. 2015.

10. Kanda T, Oba H, Toyoda K, Kitajima K, Furui S. Brain gadolinium deposition after administration of gadolinium-based contrast agents. Japanese Journal of Radiology. 2016;34(1):3-9.

11. Kanda T, Matsuda M, Oba H, Toyoda K, Furui S. Gadolinium Deposition after Contrast-enhanced MR Imaging. Radiology. 2015;277(3):924-5.

12. Han BK, Rigsby CK, Hlavacek A, Leipsic J, Nicol ED, Siegel MJ, et al. Computed Tomography Imaging in Patients with Congenital Heart Disease Part I: Rationale and Utility. An Expert Consensus Document of the Society of Cardiovascular Computed Tomography (SCCT): Endorsed by the Society of Pediatric Radiology (SPR) and the North American Society of Cardiac Imaging (NASCI). Journal of cardiovascular computed tomography. 2015;9(6):475-92.

13. Halliburton SS, Abbara S, Chen MY, Gentry R, Mahesh M, Raff GL, et al. SCCT guidelines on radiation dose and dose-optimization strategies in cardiovascular CT. Journal of cardiovascular computed tomography. 2011;5(4):198-224.

14. AAPM Task Group 23 of the Diagnostic Imaging Council CT Committee. The Measurement, Reporting, and Management of Radiation Dose in CT [AAPM Report 96]. College Park, MD: American Association of Physicists in Medicine; January 2008.

15. AAPM Task Group 204. AAPM REPORT NO. 204 Size-Specific Dose Estimates (SSDE) in Pediatric and Adult Body CT Examinations. 2011.

16. Downing TE, McDonnell A, Zhu X, Dori Y, Gillespie MJ, Rome JJ, et al. Cumulative Medical Radiation Exposure Throughout Staged Palliation of Single Ventricle Congenital Heart Disease. Pediatric cardiology. 2014.

17. Johnson JN, Hornik CP, Li JS, Benjamin DK, Jr., Yoshizumi TT, Reiman RE, et al. Cumulative radiation exposure and cancer risk estimation in children with heart disease. Circulation. 2014;130(2):161-7.

18. Glatz AC, Purrington KS, Klinger A, King AR, Hellinger J, Zhu X, et al. Cumulative exposure to medical radiation for children requiring surgery for congenital heart disease. The Journal of Pediatrics. 2014;164(4):789-94 e10.

19. Watson TG, Mah E, Schoepf UJ, King L, Huda W, Hlavacek AM. Effective radiation dose in computed tomographic angiography of the chest and diagnostic cardiac catheterization in pediatric patients. Pediatric Cardiology. 2013 Mar 1;34(3):518-24.

20. Brown DW, Gauvreau K, Powell AJ, Lang P, Colan SD, Del Nido PJ, et al. Cardiac magnetic resonance versus routine cardiac catheterization before bidirectional glenn anastomosis in infants with functional single ventricle: a prospective randomized trial. Circulation. 2007;116(23):2718-25.

21. Brown DW, Gauvreau K, Powell AJ, Lang P, del Nido PJ, Odegard KC, et al. Cardiac magnetic resonance versus routine cardiac catheterization before bidirectional Glenn anastomosis: long-term follow-up of a prospective randomized trial. The Journal of Thoracic and Cardiovascular Surgery. 2013;146(5):1172-8.

22. Han BK, Vezmar M, Lesser JR, Michalak G, Grant K, Dassenko D, et al. Selective use of cardiac computed tomography angiography: An alternative diagnostic modality before second-stage single ventricle palliation. The Journal of Thoracic and Cardiovascular Surgery. 2014;148(4):1548-54.

23. Fogel MA. Is routine cardiac catheterization necessary in the management of patients with single ventricles across staged Fontan reconstruction? No! Pediatric Cardiology. 2005;26(2):154-8.

24. Fogel MA, Pawlowski TW, Whitehead KK, Harris MA, Keller MS, Glatz AC, et al. Cardiac Magnetic Resonance and the Need for Routine Cardiac Catheterization in Single Ventricle Patients Prior to Fontan: A Comparison of 3 Groups: Pre-Fontan CMR Versus Cath Evaluation. Journal of the American College of Cardiology. 2012;60(12):1094-102.

25. Prakash A, Khan MA, Hardy R, Torres AJ, Chen JM, Gersony WM. A new diagnostic algorithm for assessment of patients with single ventricle before a Fontan operation. The Journal of Thoracic and Cardiovascular Surgery. 2009;138(4):917-23.

26. Ramamoorthy C, Haberkern CM, Bhananker SM, Domino KB, Posner KL, Campos JS, et al. Anesthesia-related cardiac arrest in children with heart disease: Data from the Pediatric Perioperative Cardiac Arrest (POCA) registry. Anesthesia and Analgesia. 2010;110(5):1376-82.

27. Girshin M, Shapiro V, Rhee A, Ginsberg S, Inchiosa MA, Jr. Increased risk of general anesthesia for high-risk patients undergoing magnetic resonance imaging. Journal of Computer Assisted Tomography. 2009;33(2):312-5.

28. Dorfman AL, Odegard KC, Powell AJ, Laussen PC, Geva T. Risk factors for adverse events during cardiovascular magnetic resonance in congenital heart disease. Journal of Cardiovascular Magnetic Resonance : Official journal of the Society for Cardiovascular Magnetic Resonance. 2007;9(5):793-8.

29. Rappaport B, Mellon RD, Simone A, Woodcock J. Defining safe use of anesthesia in children. The New England Journal of Medicine. 2011;364(15):1387-90.

30. Hays SR, Deshpande JK. Newly postulated neurodevelopmental risks of pediatric anesthesia. Curr Neurol Neurosci Rep. 2011;11(2):205-10.

31. Wilder RT, Flick RP, Sprung J, Katusic SK, Barbaresi WJ, Mickelson C, et al. Early exposure to anesthesia and learning disabilities in a population-based birth cohort. Anesthesiology. 2009;110(4):796-804.

32. Flick RP, Katusic SK, Colligan RC, Wilder RT, Voigt RG, Olson MD, et al. Cognitive and behavioral outcomes after early exposure to anesthesia and surgery. Pediatrics. 2011;128(5):e1053-61.

33. DiMaggio C, Sun LS, Kakavouli A, Byrne MW, Li G. A retrospective cohort study of the association of anesthesia and hernia repair surgery with behavioral and developmental disorders in young children. J Neurosurg Anesthesiol. 2009;21(4):286-91.

34. Fogel MA, Weinberg PM, Parave E, Harris C, Montenegro L, Harris MA, et al. Deep sedation for cardiac magnetic resonance imaging: A comparison with cardiac anesthesia. The Journal of Pediatrics. 2008;152(4):534-9, 9 e1.

35. Odegard KC, DiNardo JA, Kussman BD, Shukla A, Harrington J, Casta A, et al. The frequency of anesthesia-related cardiac arrests in patients with congenital heart disease undergoing cardiac surgery. Anesthesia and Analgesia. 2007;105(2):335-43.

36. Miller JH, Hu HH, Pokorney A, Cornejo P, Towbin R. MRI Brain Signal Intensity Changes of a Child During the Course of 35 Gadolinium Contrast Examinations. Pediatrics. 2015;136(6):e1637-40.

37. Kanda T, Oba H, Toyoda K, Furui S. Recent Advances in Understanding Gadolinium Retention in the Brain. AJNR American Journal of Neuroradiology. 2016;37(1):E1-2.

38. Roberts DR, Holden KR. Progressive increase of T1 signal intensity in the dentate nucleus and globus pallidus on unenhanced T1-weighted MR images in the pediatric brain exposed to multiple doses of gadolinium contrast. Brain & Development. 2016;38(3):331-6.

39. Kanda T, Fukusato T, Matsuda M, Toyoda K, Oba H, Kotoku J, et al. Gadolinium-based Contrast Agent Accumulates in the Brain Even in Subjects without Severe Renal Dysfunction: Evaluation of Autopsy Brain Specimens with Inductively Coupled Plasma Mass Spectroscopy. Radiology. 2015;276(1):228-32.

40. Han BK, Lesser JR. CT imaging in congenital heart disease: an approach to imaging and interpreting complex lesions after surgical intervention for tetralogy of Fallot, transposition of the great arteries, and single ventricle heart disease. Journal of Cardiovascular Computed Tomography. 2013;7(6):338-53.

41. Khairy P, Van Hare GF, Balaji S, Berul CI, Cecchin F, Cohen MI, et al. PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease. Heart rhythm : The official journal of the Heart Rhythm Society. 2014.

42. Guo YK, Gao HL, Zhang XC, Wang QL, Yang ZG, Ma ES. Accuracy and reproducibility of assessing right ventricular function with 64-section multi-detector row CT: Comparison with magnetic resonance imaging. International Journal of Cardiology. 2010;139(3):254-62.

43. van der Vleuten PA, de Jonge GJ, Lubbers DD, Tio RA, Willems TP, Oudkerk M, et al. Evaluation of global left ventricular function assessment by dual-source computed tomography compared with MRI. European Radiology. 2009;19(2):271-7.

44. Groen JM, van der Vleuten PA, Greuter MJ, Zijlstra F, Oudkerk M. Comparison of MRI, 64-slice MDCT and DSCT in assessing functional cardiac parameters of a moving heart phantom. European Radiology. 2009;19(3):577-83.

45. Plumhans C, Muhlenbruch G, Rapaee A, Sim KH, Seyfarth T, Gunther RW, et al. Assessment of global right ventricular function on 64-MDCT compared with MRI. American Journal of Roentgenology. 2008;190(5):1358-61.