

Late Complications and Sequelae in Patients after Congenital Heart Disease Operation



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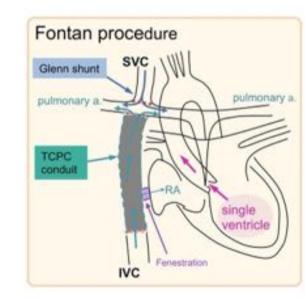
> ☑ The author has no conflict of interest to disclose with respect to this presentation.

Introduction

Recently patients with congenital heart disease (CHD) can survive long term thanks to the advancement of surgical procedure and subsequent medical treatment, and diagnostic radiologists encounter the images of these patients with increasing frequency. As most of the patients present with residua, sequelae or complications, radiologists need to have enough knowledge of

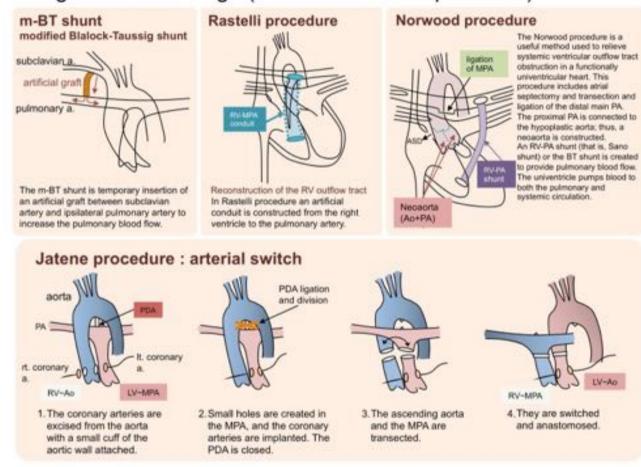
In this presentation we will especially focus on various image findings of late complications and sequelae in the patients and consider what is valuable information and suggestion to clinicians.

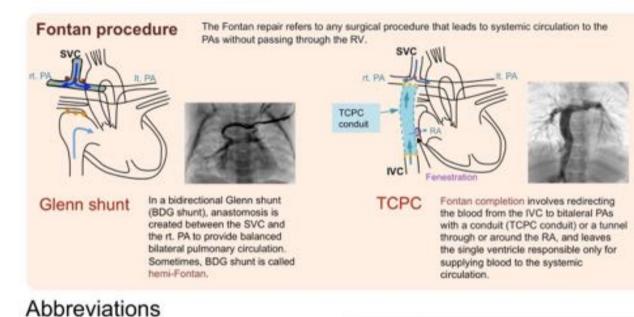
Especially, long-term management in the CHD patients after Fontan procedure is not yet satisfactory. The hemodynamics after Fontan procedure is quite different from normal biventricular circulation, and late complications are major problems; so-called post-Fontan syndrome.



The Fontan procedure is a palliative operation performed in patients with a functional or anatomic single ventricle, that refers to any operation that results in the flow of systemic venous blood to the pulmonary artery without passing through a ventricle, and the single ventricle provides systemic blood flow. Fontan circulation is characterized by high central venous pressure and low cardiac output, that progressively decrease function of all organ systems. Liver dysfunction, protein-losing enteropathy, plastic bronchitis, collateral vessels are often observed in the follow-up period after CHD operation, especially Fontan procedure.

Background Knowledge (Schema of CHD operations)





Atrial septal defect cTGA

Atrioventricular septal defect Patent ductus arteriosus Pulmonary stenosis Coarctation of the aorta rt. AoA Complete transposition of the great arteries right aortic arch Double inlet left ventricle Single ventricle Damus-Kaye-Stansel procedure Tricuspid atresia Double outlet right ventricle Truncus arteriosus communis Total anomalous pulmonary venous return Hypoplstiac left heart syndrome TCPC MAPCA Major aortopulmonary collateral artery Total cavopulmonary connection m-BT shunt modified Blalock-Taussig shunt Tetralogy of Fallot Pulmonary atresia Ventricular septal defect Contents

Pulmonary atresia with intact ventricular septum

A. Cardiac insufficiency

 ✓ Cardiac enlargement B. Liver dysfunction Passive hepatic congestion ✓ Multiple nodules in cirrhotic liver

D. Plastic bronchitis

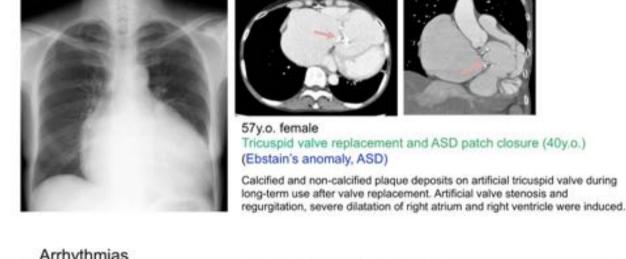
- ✓ Hepatocellular carcinoma (HCC) in cirrhotic liver ✓ Amiodarone hepatotoxity C. Protein-losing enteropathy (PLE)
- E. Collateral vessels ✓ Pulmonary arteriovenous malformations (PAVMs) ✓ Veno-venous collaterals (Veno-venous shunts; VV shunts) ✓ Aortopulmonary collaterals (APCs)
- F. Coagulation abnormalities ✓ Thrombus in the conduit, Thrombus in right ventricle ✓ Cerebral infarction
- ✓ Intestinal ischemia ✓ Myocardial infarction
- G. Organ atrophy ✓ Brain atrophy, Transient brain atrophy
- H. Bleeding tendency ✓ Intracranial hemorrhage ✓ Massive ovarian hemorrhage
- ✓ Pseudoaneurysm
- Gallstone ✓ Bilirubin stone ✓ Ceftriaxone associated biliary pseudolithiasis
- J. Infections ✓ Infective endocarditis (Vegetations) ✓ Brain abscess Lung abscess
- K. Pulmonary congestion, Lung damage L. Diaphragmatic paralysis M. Aortic insufficiency ✓ Dilatation of ascending aorta

✓ Septic pulmonary embolism

- ✓ Aortic dissection N. Others
- ✓ Blood flow stagnation ✓ Residual shunts ✓ Coronary fistulas
- ✓ Ventricular aneurysm, Stenosis of conduit Deformities of vessels and bronchi ✓ Relative narrow conduit due to the patient's growth
- Degradation of prosthetic materials ✓ Arteriovenous fistula at the puncture site. Localized aortic dissection and stenosis after balloon dilatation ✓ Vessel occlusion
- ✓ Anastomotic stenosis ✓ Thoracic deformity
- Cardiac insufficiency

Cardiac enlargement

Multiple mechanisms induce cardiac insufficiency in postoperative CHD patients, which involve too much enlargement of ventricle, inability of the systemic ventricle, pressure overload, blood flow imbalance, abnormalities of myocardial tissue and deformity of thoracic cage.

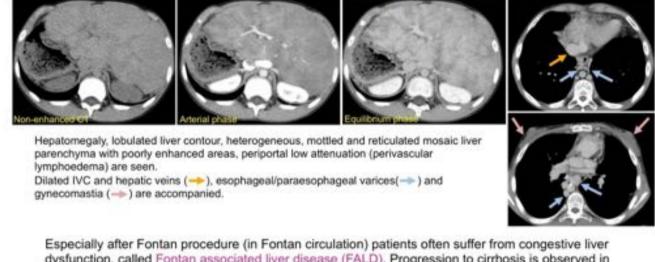


Arrhythmias are the most common long-term complications in patients after CHD operation, and which increase with aging and lesion complexity. All types of atrial and ventricular arrhythmias occur, and multiple types often coexist. The cause lies in injuries of the sinus node or atrium during operation. The internodal conduction pathways are interrupted, and reentry circuits are formed. Especially patients after classic Fontan procedure (atriopulmonary connection) have a risk of atrial arrhythmias due to severely enlarged right atrium. Atrial arrhythmia was caused due to blood stasis, increased blood viscosity, and leads to thromboembolic complications. Thus, anticoagulation therapy is recommended for patients with atrial arrhythmias. The treatment of arrhythmias includes antiarrhythmic drug therapy (Amiodarone and β-blockers), electrical cardioversion, implantation of a pacemaker and implantable cardioverter defibrillator (ICD).

B. Liver dysfunction

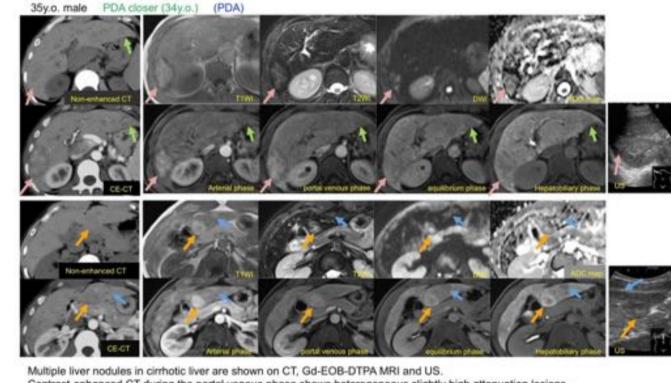
Postoperative CHD patients are placed under conditions with such as long-term hypoxia, low cardiac output, elevated central venous pressure and multiple microthrombosis. Chronic congestive heart increases hepatic venous pressure, which leads to congestive hepatopathy, portal venous hypertension. Furthermore, portal hypertension leads to splenomegaly and portosystemic shunts. Liver dysfunction causes coagulopathy, immunodeficiency and development of liver cirrhosis. Patients with liver cirrhosis have a risk of hepatocellular

Passive hepatic congestion 19y.o. male Fontan procedure (extracardiac TCPC: 3y7m) (Asplenia, AVSD, Single RV, PS, TAPVR)



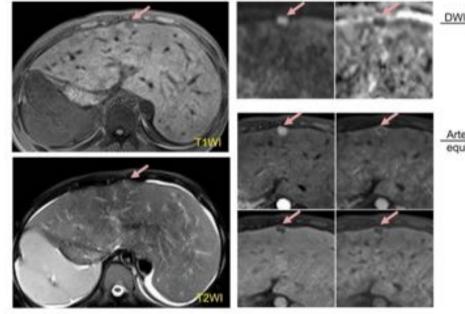
dysfunction, called Fontan associated liver disease (FALD). Progression to cirrhosis is observed in patients within 10 years after the initial Fontan procedure.

Multiple regenerative nodules in cirrhotic liver



Contrast-enhanced CT during the portal venous phase shows heterogeneous slightly high attenuation lesions. MRI shows multiple lesions, which are moderately high intense on T1WI, high intense on T2WI, DWI and ADC map. Arterial phase of enhanced MRI shows enhancing lesions, and hepatobiliary phase shows moderately high, iso or moderately low intense regenerative nodules.

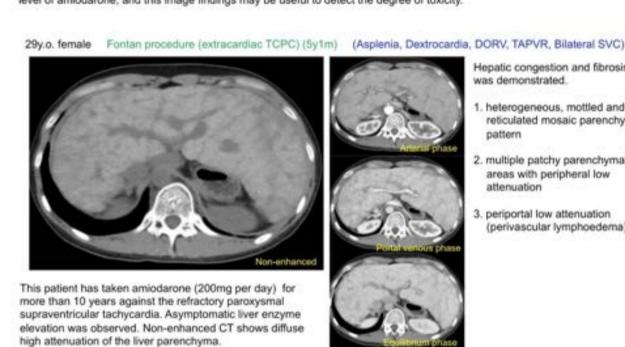
Hepatocellular carcinoma (HCC) in cirrhotic liver 29y.o. female Fontan procedure (intracardiac TCPC) (10y.o.) (Heterotaxy (Polysplenia), Single ventricle, TGA, AVSD)



Liver cirrhosis with nodular surface, ascites and splenomegaly are shown on MRI. MRI reveals a nodule in diameter of 15 mm with moderately low intensity on T1WI, moderately high intensity on T2WI, restricted diffusion on DWI and ADC map, homogeneous arterial enhancement, and completely washout on the hepatobiliary phase at S4 of the liver. This nodule was finally diagnosed as HCC.

Amiodarone hepatotoxity

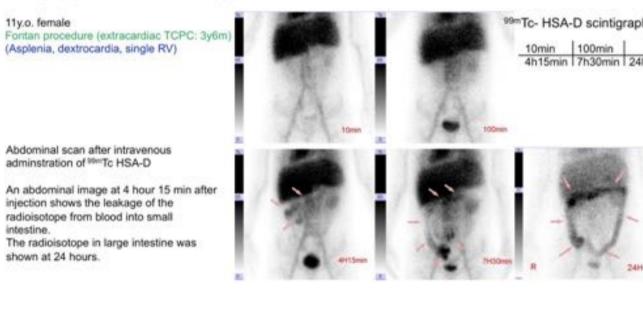
Amiodarone is an iodine-containing antiarrhythmic drug. Amiodarone is a highly effective treatment for supraventricular and ventricular tachyarrhythmia. Long-term administration of this drug causes hepatotoxicity due to iodine accumulation in the liver. In addition, the lungs, myocardium, thyroid, spleen and pancreas are also iodine deposited organs. Amiodarone could be associated with several serious adverse effects. Amiodarone hepatotoxicity is characterized by histologic steatosis, inflammation, fibrosis, and phospholipidosis, and it may progress to cirrhosis and result in decompensated hepatic failure. CT shows diffuse high attenuation of the liver parenchyma, which is depend on iodine concentration in liver and reflects the tissue level of amiodarone, and this image findings may be useful to detect the degree of toxicity.



Hepatic congestion and fibrosis heterogeneous, mottled and nultiple patchy parenchymal areas with peripheral low attenuation periportal low attenuation perivascular lymphoedema)

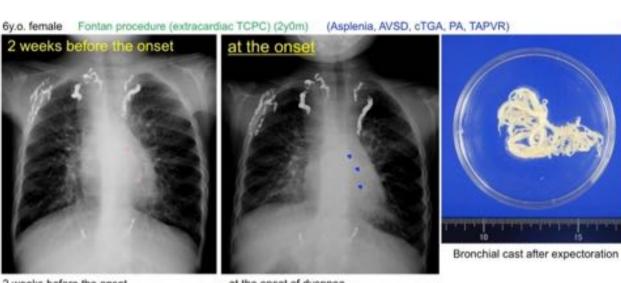
Protein-losing enteropathy (PLE) Protein-losing enteropathy (PLE) is defined as excessive loss of proteins across the intestinal mucosa, and that leads to refractory ascites, generalized edema and pleural effusion. PLE is rare in the early years after

CHD operation, and increases with time progress. The prevalence of Fontan-associated PLE is 11.1%. The etiology of PLE is unknown. However, PLE is considered to be caused by high systemic venous pressure, low cardiac output and high pulmonary vascular resistance. PLE often leads to hypoproteinemia, immunodeficiency, hypocalcemia, and coagulopathy. The prognosis is poor, and about half of the patients Technetium 99m-diethylenetriamine penta-acetic acid-labeled human serum albumin (99mTc HSA-D) scintigraphy is the most valuable diagnostic method.



Plastic bronchitis Plastic bronchitis is a rare but serious complication of the Fontan procedure, occurring in less than 1-2% of

the patients. It is characterized as noninflammatory mucinous casts that form in the tracheobronchial tree Although the etiology is unclear, it is considered to be associated with high central venous pressure, and multiple lymphatic collateral vessels lead to the development of lymphoalveolar fistula and bronchial casts.



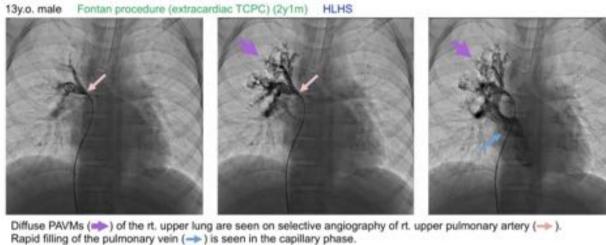
2 weeks before the onset at the onset of dyspnea Lt. lower bronchus is not visible due to Lt. lower bronchus is visible. the presence of bronchial cast. Coil embolization of APCs (aortopulmonary collaterals) from bilateral intercostal arteries.

Collateral vessels

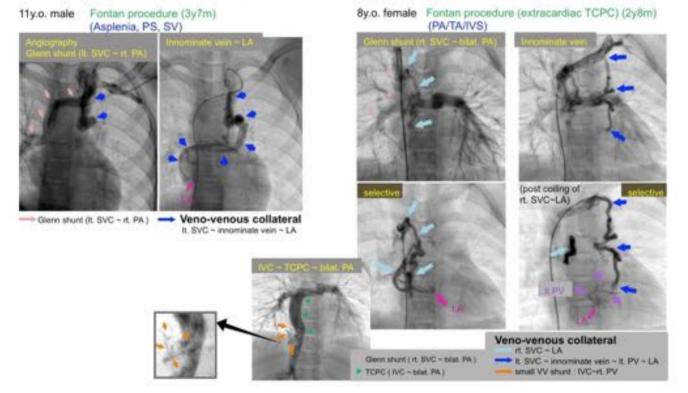
Pulmonary arteriovenous malformations Pulmonary arteriovenous malformations (PAVMs) are structurally abnormal vessels that provide direct capillary-free communication between pulmonary artery and vein. PAVMs is developed by the unbalanced pulmonary perfusion of

rt. subscapular artery and bilateral subphrenic arteries are also seen on chest X-ray images

hepatic venous blood flow after CHD operation (especially Fontan procedure) and seen in the affected lungs with a lack of hepatic venous blood flow. Decreased hepatic venous return results in abnormalities in angiogenesis. Precursors of angiogenesis inhibitors are produced in liver, and a reduced exposure of angiogenesis inhibitors in pulmonary vessels is considered to be the main cause of PAVMs.

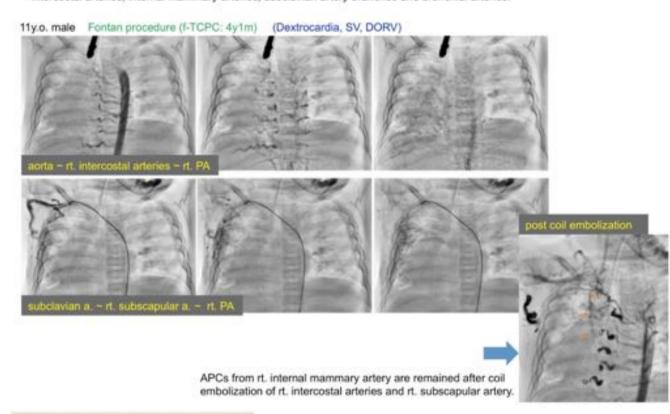


Veno-venous collaterals (Veno-venous shunts ; VV shunts) (Communication between systemic vein and pulmonary vein or left atrium) Veno-venous collaterals occur in patients with chronically elevated systemic venous pressure, such as the Fontan circulation. These collaterals result in connections from the systemic veins (vena cava or innominate vein) to the pulmonary veins or to the left atrium. These collaterals act as a right-to-left shunt, causing systemic desaturation, contributing to the polycythemia, and also posing a risk for paradoxical embolism. These can be treated with transcatheter embolization, but often re-grow.



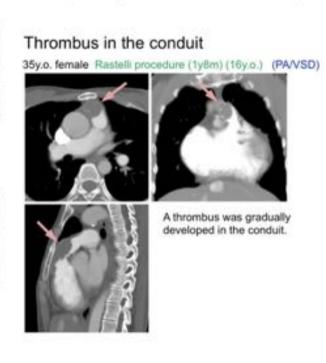
Aortopulmonary collaterals

Aortopulmonary collaterals (APCs) is a left-to-right shunt. Hypoxia is a potent stimulus for the development of APCs. These collaterals supply blood from the systemic arteries to the pulmonary circulation, and may results in volume overload of the systemic ventricle, increased pulmonary blood flow and pressure. APCs typically arise from the descending aorta, intercostal arteries, internal mammary arteries, subclavian artery branches and bronchial arteries.



Coagulation abnormalities

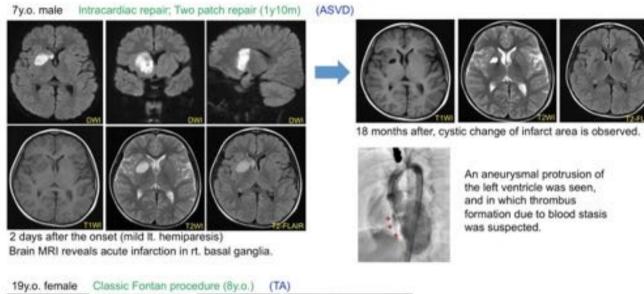
Coagulation abnormalities are highly prevalent after CHD operation. The cause is multifactorial (abnormal venous flow, low cardiac output, the lack of AV synchrony, polycythemia due to chronic cyanosis, and abnormalities of the coagulation cascade, multiple suture sites, prosthetic materials, blind cavities). Thromboembolic events (pulmonary embolism, cerebral infarction, and so on) are often observed. Silent pulmonary thromboemboli have been reported in 17% of adult patients following Fontan procedure.



Thrombus in right ventricle function due to tricuspid atresia, and thrombus is formed in the

Cerebral infarction

Atrial arrhythmias, blood stasis and increased blood viscosity may lead to intraatrial and intraventricular thrombus and ischemia of multiple organs. Cerebral infarction rates in postoperative CHD patients have been reported as 9 to 12 times higher (<55y.o.) and 2 to 4 times higher (55-64y.o.) compared to the general population.



Multiple old infarcts in It, cerebral white matter accompanying brain atrophy are shown. MRA shows severely narrowed distal branch of It. MCA.

Intestinal ischemia



Contrast-enhanced CT shows

cerebral blood flow in the

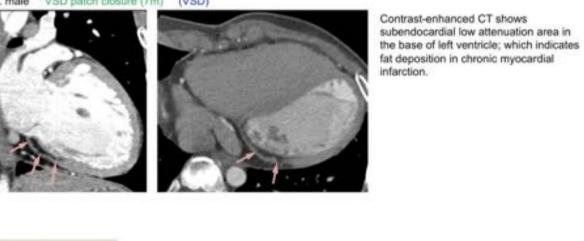
second Fontan operation.

trast-enhanced CT shows circumferential wall

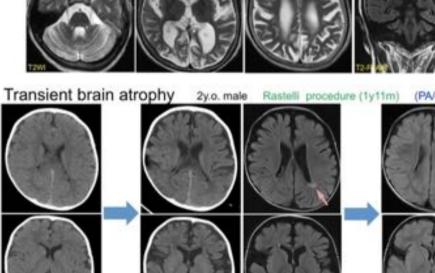
ickening and submucosal edema involving the

microembolism were considered to be the cause of

ascending colon. Poor cardiac output and



G. Organ atrophy Atrophy of organs may be induced by hypoxia due to intraoperative low blood pressure or chronic low blood flow. Brain atrophy 25v.o. female Fontan procedure (6y.o., 13y.o.) (SV, Asplenia, persistent it. SVC)

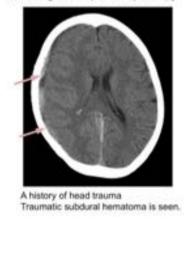


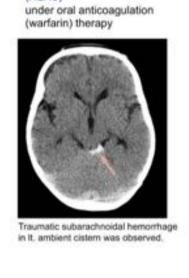
T at 17 days and MRI at 44 days after Rastelli procedure show brain atrophy in bilateral ontal and temporal lobes. On MRI at 44 days, a periventricular hyper-intense esion appeared. One year after, these malities were obscured. This image findings may be associated with transient cerebral ischemia during surgery and recovery.

. Bleeding tendency In postoperative CHD patients with cyanosis, thrombocytopenia, abnormality of platelet function, decrease

of coagulation factors are often observed. Moreover, vascular shear stress increases when red blood cells increase, and the increased production of nitric oxide and prostaglandin induces arteriolar dilatation and the growth of capillary vessels. In addition, these patients often take anticoagulants or antiplatelet agents. These factors leads to bleeding tendency.

Traumatic subdural hematoma Subarachnoidal hemorrhage Cerebral hemorrhage Classic Fontan procedure (8y6m) Fontan procedure (2y5m) Fontan procedure (4v8m) (Dextrocardia, SV, PS) under long-term antiplatelet (low under oral anticoagulation under long-term oral (warfarin) therapy dose aspirin) therapy anticoagulation (warfarin) therapy







Massive ovarian haemorrhage 22y.o. female VSD/ASD closure, CoA repair, Mitral valve replacement using mechanical valve (5y5m) (VSD, ASD, CoA) under long-term oral anticoagulation (warfarin) therapy

This patient presented to the emergency department with abdominal pain and vomiting at 21 days after menstruation (Luteal phase). Huge ovarian bleeding (intraabdominal hemorrhage) was seen. Pseudoaneurysm 11y.o. female Fontan procedure (2y6m) (PA/IVS, HypoRV)



Gallstone Gallstone is one of the well-known major complications in postoperative CHD patients with cyanosis. Cyanosis

induces secondary erythrocytosis to maintain adequate tissue oxygen delivery. Thus, increasing heme breakdown and bilirubin production is a major factor in the pathogenesis of gallstones. Almost 1% of the patients with cyanotic CHD have gallstone in Japan.

Ceftriaxone associated biliary pseudolithiasis Approximately 40% of ceftriaxone is excreted into the bile and concentrated in the gallbladder. High concentration of ceftriaxone-calcium complex in gallbladder induce gallstone formation.



She presented with vomiting. Elevated serum bilirubin level was observed.

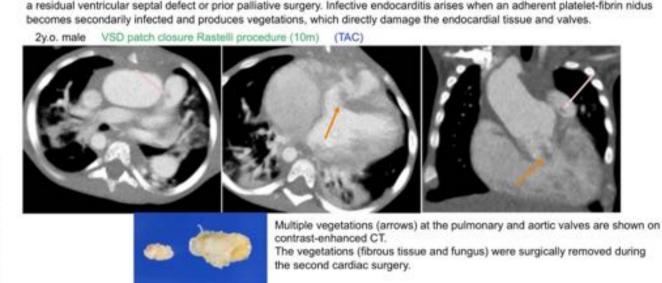
Gallstones and CBD stones were seen.

He complained of right hypochondralgia and vomiting after the administration of antibiotics (ceftriaxone 120 mg/kg/day) for 6 weeks with suspect of infective endocarditis. CT showed gallstones and CBD stones. Finally ceftriaxone associated biliary pseudolithiasis was diagnosed.

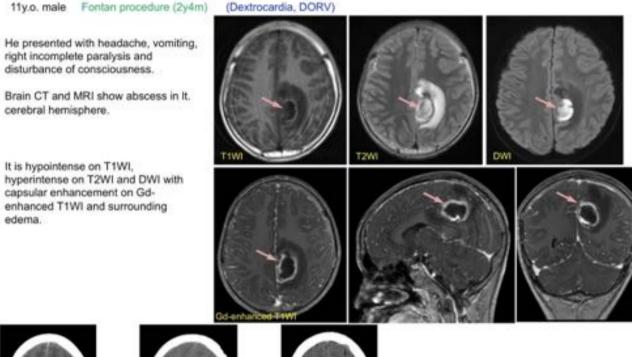
Infections

Right-to-left shunt does not pass through the pulmonary circulation, where bacteria are intercepted by phagocytosis. This allows direct entry of the bacterial nest to systemic circulation. Additionally, polycythemia and organ hypoxemia create favourable circumstances for the development of abscess. Systemic infections such as pneumonia, lung abscess, infective endocarditis, brain abscess are frequent in patients after CHD

Infective endocarditis (Vegetations) Patients with prosthetic valves are especially prone to endocarditis. The risk of infective endocarditis is very low when the postoperative CHD patient has a left-to-right shunt or valve stenosis; but it increases in the patients with complex cyanotic CHD,



Brain abscess



Orilling drainage was performed, 10cc

yellowish gray liquid with irritating

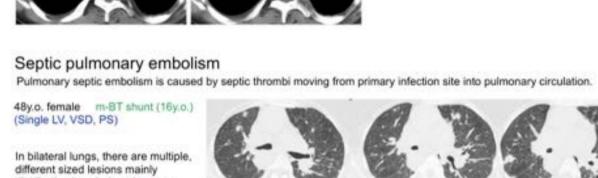
Abscess formation in rt. lung is more obvious on

risk of pulmonary infection due to hypo-perfusion

ostoperative CHD patients with cyanosis have a

ontrast-enhanced CT.

Lung abscess



distributing in subpleural areas. Small abscess cavities (----) and demonstrated

K. Pulmonary congestion, Lung damage

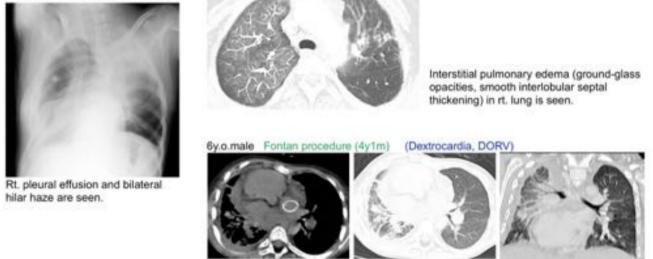
Pleural effusion

19y.o.female

In postoperative CHD patients, lymphatic circulation may be affected by high systemic venous pressure and impaired thoracic duct flow. Increased pulmonary lymphatic pressure results in interstitial pulmonary edema or lymphedema, and leakage into the thorax or pericardium leads to pericardial and pleural effusions. Moreover, chylothorax is caused by injury of the thoracic duct, venous or lymphatic congestion, central vein thrombosis, or diffuse injury of mediastinal lymphatic tissue. Chylothorax is particularly frequent in patients with the Glenn shunt and Fontan procedures.

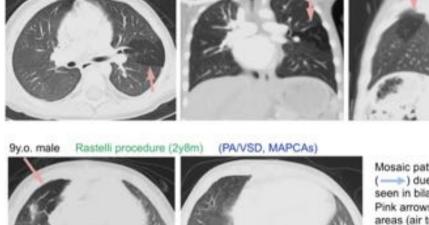
25y.o.female Glenn shunt (6y1m) (PA/IVS)

Pulmonary congestion



Lung damage Lung damage commonly occurs due to pulmonary congestion, diaphragmatic injury, diaphragmatic paralysis, restrictive thoracic cage after thoracotomy/stemotomy, abnormalities in lung parenchyma, lung hypoplasia from altered blood flow, drug toxicity, and so on. Restrictive lung disease is especially common in patients who have undergone multiple

vessels and cardiomegaly are seen.

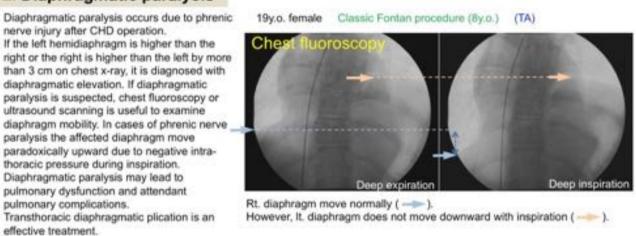


Mosaic pattern of attenuation and atelectasis ->) due to small air way obstruction are Pink arrows (-----) indicate lower attenuation areas (air trapping).

trapping) due to bronchiolar collapse in It. upper lung is

.. Diaphragmatic paralysis

nerve injury after CHD operation. If the left hemidiaphragm is higher than the right or the right is higher than the left by more than 3 cm on chest x-ray, it is diagnosed with diaphragmatic elevation. If diaphragmatic paralysis is suspected, chest fluoroscopy or ultrasound scanning is useful to examine diaphragm mobility. In cases of phrenic nerve paralysis the affected diaphragm move paradoxically upward due to negative intrahoracic pressure during inspiration. Diaphragmatic paralysis may lead to pulmonary dysfunction and attendant Fransthoracic diaphragmatic plication is an effective treatment.



Atelectasis associated with diaphragmatic paralysis



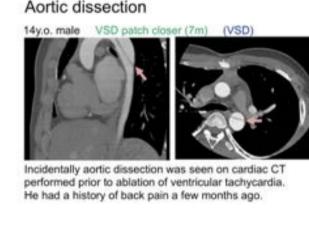
root and valvular deformation resulting from retraction of the surgical patch. Patients after DKS procedure or Jatene procedure (arterial switch) may have neoaortic dilation. Aortic dilatation may lead to dissection or rupture; however it is rare.

Some patients develop progressive aortic insufficiency. Aortopathy (abnormalities in the aortic medial layer,

aortic regurgitation and aortic and ventricular dysfunction) is mainly caused by the dilatation of the aortic

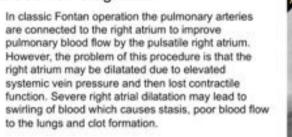


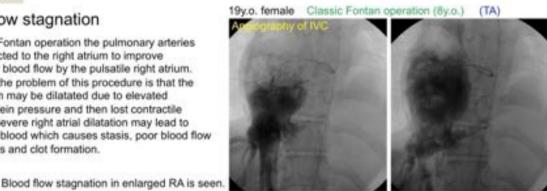
Dilatation of ascending aorta



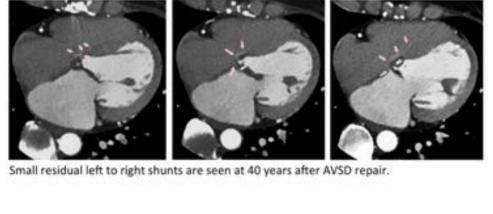
N. Others Blood flow stagnation

Enlargement of the ascending aorta is seen.

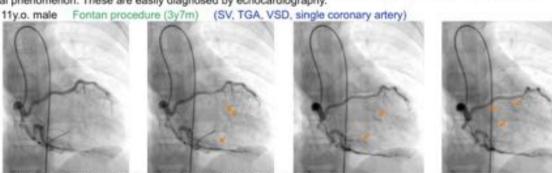




Residual shunts In some cases, residual shunts may remain hemodynamically and clinically irrelevant. If postsurgical residual VSDs are less than 2 mm, they are expected to be closed spontaneously in the majority within a year.

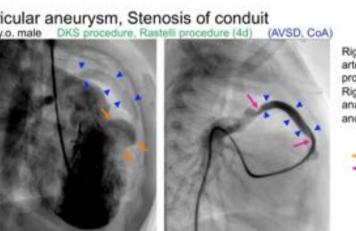


Coronary fistula may occurr after myocardial resection. These are too small to cause cyanosis, but rarely cause coronary steal phenomenon. These are easily diagnosed by echocardiography.



Ventricular aneurysm, Stenosis of conduit

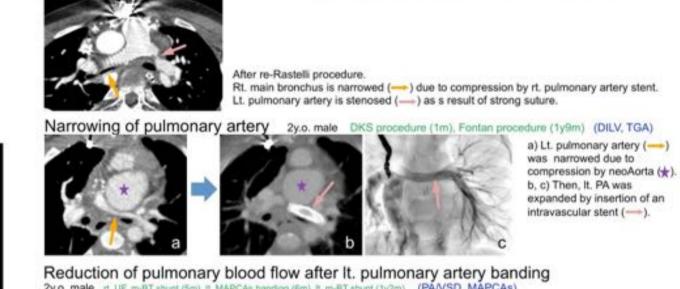
Coronary angiogram reveals multiple fistulas of coronary artery.



Right ventricle and bilateral pulmonary arteries are connected by a conduit (Rastelli procedure) (>). Right ventricular aneurysm near the conduit anastomosis and stenoses at the proximal and distal sites of the conduit are seen. Right ventricular aneurysm

Deformities

Narrowing of bronchus 11y.o. male Rastelli procedure (2y.o.) (11y.o.) (PAVSD, MAPCAs, rt. AoA)



After It. MAPCAs were Lt. subclavian artery was connected banded(), blood flow of It. to it. pulmonary artery (m-BT shunt)



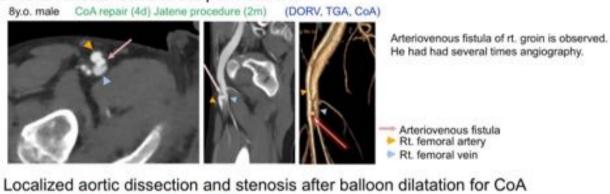
required before adulthood. he conduit is relatively narrow and a larger sized extended due to his The blood flow from RV to PA was

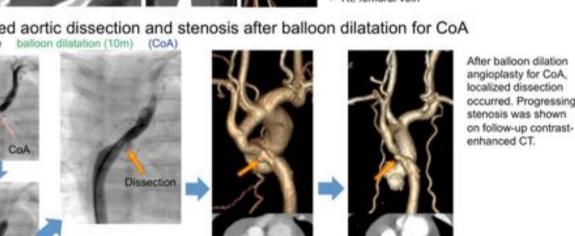


Arteriovenous fistula at the puncture site

Degradation of prosthetic materials

Rastelli procedure is

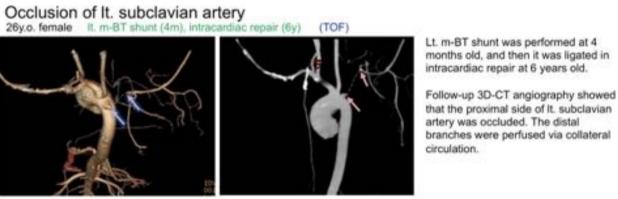




Vessel occlusion

1v.o. male





Anastomotic stenosis

Conclusion



After negAorta (*) had been constructed, anastomotic stenosis (----) was observed. Balloon dilation angioplasty was repeatedly performed. Mild stenosis of distal aortic arch (CoA) (----) is also seen

Femoral artery flow (4) perfused

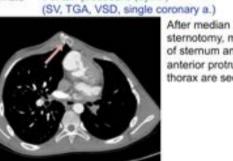
ventricle is compressed

by the concave

deformed thorax.

via collateral vessels (----)

Repeated open thoracic surgery may result in thoracic deformity, and which may accompany organ compression. 16y.o. male Fontan procedure (3y7m)



in postoperative CHD patients.

of sternum and interior protrusion of



Thank you for visiting our presentation.

We presented some image findings of late complications and sequelae

We think that this presentation provide valuable knowledge to clinical

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