

HISTORY

Experimental work (Carlon, Mondini and deMarchi, 1951, Glenn and Patino, 1954) explored the feasibility of creating a connection between the superior vena cava (SVC) and the pulmonary artery to increase pulmonary blood flow. William W.L. Glenn, a professor of surgery at Yale University, first reported the clinical application of this concept in 1958.

INDICATIONS

Since the late 1950's the Glenn shunt, as the SVC-to-right pulmonary artery anastomosis is called, has been performed on patients with diverse cyanotic congenital heart disease to improve pulmonary blood flow. The Glenn shunt does not create volume overload of the ventricle or increased work for the ventricle, as is the case in systemic-pulmonary artery shunts. It provides venous flow to the lung fields for oxygenation, rather than an arteriovenous mixture. The venous return is under relatively low pressure, unlike systemic-pulmonary artery shunts, and the risk for pulmonary artery distortion and late pulmonary vascular obstructive disease is substantially less.

The Glenn shunt is palliative, not corrective. Depending on the diagnosis and the surgical era, the Glenn shunt may be the only palliation for the cyanotic patient, one of several palliative surgeries, or a step prior to corrective surgery or the Fontan form of total right heart bypass.

CLASSIC GLENN SHUNT

Dr. Glenn described an anastomosis between the transected distal end of the right pulmonary artery and the side of the SVC, which is ligated distal to the anastomosis. The azygous vein is ligated to prevent its decompressing flow from the SVC. Systemic venous return from the head and upper extremities is to the right lung, driven by the pressure gradient from the SVC to the left atrium.

A patient having total right heart bypass for palliation of cyanotic congenital heart disease with a Fontan procedure after a classic Glenn shunt typically would have an anastomosis between the inferior vena cava (IVC) and the left lung, using either a direct atrio-pulmonary connection or a conduit. Due to requisite extensive dissection and technically difficult pulmonary artery reconstruction, patients after a classic Glenn usually do not receive extra-cardiac IVC-pulmonary artery conduits as are now commonly performed in the Fontan type procedure.

BI-DIRECTIONAL CAVO-PULMONARY SHUNT

The bi-directional cavo-pulmonary shunt was first performed in 1966. The transected end of the SVC is anastomosed to the side of the undivided right pulmonary artery, allowing flow to both lung fields. As with the classic Glenn shunt, the bi-directional cavo-pulmonary shunt is far less likely to engender pulmonary vascular obstructive disease compared with systemic-pulmonary shunts, and there is minimal distortion of the pulmonary artery architecture. With newer surgical techniques the bi-directional Glenn (as it is sometimes called) can be performed without cardiopulmonary bypass. If bilateral vena cavae exist, both can be anastomosed end-to-side to the pulmonary artery. A bi-directional cavo-pulmonary shunt may be done under six months of age, assuming the pulmonary vascular resistance has declined.

The bi-directional cavo-pulmonary shunt currently most commonly is employed as the first stage in a staged Fontan procedure. (Some surgeons feel that the staged Fontan improves results. Others feel a fenestration in the Fontan circuit at the time of a single Fontan operation with subsequent closure of the fenestration improves results—by improving cardiac output, minimizing systemic venous hypertension, and decreasing post-operative thoracostomy drainage—at the cost of relative desaturation until the fenestration is closed.) It may serve as definitive palliation if the Fontan is thought too risky. In this setting and depending on the specific cyanotic condition, the bi-directional cavo-pulmonary shunt can provide a 1 _ ventricular repair; increased bilateral pulmonary blood flow is achieved yet ventricular work and volume is not increased as is true with systemic-pulmonary shunts.

Aortopulmonary collaterals may occur after a bi-directional cavo-pulmonary anastomosis, providing competitive pulsatile pulmonary blood flow. The prevalence may be as high as 36% in patients after a bi-directional cavo-pulmonary anastomosis. Most aortopulmonary collateral vessels originate from the internal mammary artery or thyrocervical trunk. The significance these vessels and indications for closure are not yet clear.

OUTCOME

Operative mortality

In a series from Toronto the 30-day operative mortality rate after a Glenn shunt was 9.6%. Death in the perioperative period is most commonly due to SVC syndrome, persistence cyanosis and cerebral injury. Poorer results are seen in patients with increased pulmonary vascular resistance, hypoplastic pulmonary arteries, or age younger than 18 months.

Long-term mortality

During follow-up in a series from Toronto the death rate was 20%. The majority of these deaths were associated with subsequent repair procedures.

Complications

Complications in the perioperative period include

- SVC syndrome, with excessive venous pressure from connection of systemic venous return into the pulmonary circulation causing symptoms.
- cerebral edema, again due to excessive venous pressure. A patent azygous vein would allow decompression of the elevated pressures in the SVC—this would, however, decrease flow down the Glenn shunt, promoting stasis and thrombosis.
- hemothoraces
- chylothoraces
- pulmonary artery thrombosis, possibly with infarction
- sinus node injury with subsequent rhythm disturbance.
- decreased flow down the Glenn shunt leading to thrombosis if bilateral SVCs are present and the left SVC is not ligated at operation.

Later complications include protein-losing enteropathy (PLE). Higher pressures in the SVC and thoracic duct can be transmitted to intestinal lymphatics, with gut loss of proteins and subsequent abnormal fluid homeostasis. PLE is more common after a Fontan operation but has been reported after a Glenn shunt.

The Glenn shunt can be taken down but with difficulty at subsequent surgery.

Mechanical ventilation in patients with a cavo-pulmonary anastomosis is challenging. Flow from SVC to left atrium is non-pulsatile, down a pressure gradient. Even small amounts of positive end-expiratory pressure (PEEP) can decrease substantially the cardiac output. It is recommended that patients have continuous negative and extra thoracic pressure instead.

Long-term efficacy

Investigators at Yale University continue to define the long-term outcomes of the Glenn shunt. The Glenn shunt is functional (that is, it provides some form of palliation with or without further corrective or palliative procedures) at 10 years in 81% of patients, at 20 years in 50% of patients. Only a minority of patients, however, can expect to be free of further operative procedures to augment pulmonary blood flow by 25 years after the Glenn shunt.

Investigators at Toronto and elsewhere corroborate that the palliation achieved by the Glenn shunt alone is adequate for 5-10 years in most patients. Thereafter further intervention is usually necessary.

The palliation from the Glenn shunt can be augmented by the surgical creation of an arteriovenous fistula, either axillary or brachial, bolstering the flow down the SVC. The flow to the contralateral lung can be increased if feasible based on the specific anatomy of the cyanotic heart condition.

The causes of worsening oxygenation late after a Glenn shunt include

- decreased flow to the contralateral lung due to progression of the specific cyanotic pathophysiology (e.g. increasing pulmonary stenosis—whereas previously antegrade flow through the pulmonary artery to the contralateral lung provided some pulmonary blood flow).
- intravascular pulmonary thrombosis.
- development of venous collateral flow from the SVC to the IVC resulting in decreased flow down the Glenn shunt to the ipsilateral lung. If a bi-directional cavo-pulmonary anastomosis is present, the collateral from the SVC into the IVC drains into chambers leading to the systemic circulation,

- resulting in a right to left shunt. These veno-venous collaterals may occur in up to 33% of patients.
- recanalization of the ligated connection between the SVC and the right atrium—again resulting in decreased flow down the Glenn shunt and, if a bi-directional cavo-pulmonary anastomosis is present, a right to left shunt. This SVC to right atrium communication can be occluded with an interventional catheterization procedure.
 - thrombosis and stricture of the Glenn shunt. Thrombosis and stricture may contribute to some cases of late cardiac death after the Glenn shunt.
 - increased pulmonary vascular resistance--primarily or as a result of increasing blood viscosity due to hypoxemia from the above causes
 - pulmonary arteriovenous malformations (AVMs). Pulmonary AVMs may be the main reason of late clinical deterioration in patients after the Glenn.
 - pulmonary ventilation-perfusion abnormalities. The classic Glenn shunt results in selective perfusion of the right lower lobe, with flow susceptible to gravitational effects. The flow in the right upper and right middle lobe is minimal. After a bi-directional cavo-pulmonary anastomosis blood flows preferentially to both lower lobes.
 - SVC drainage as a percentage of systemic venous drainage is inversely proportional to the age and size of a patient. Somatic growth with a relative decrease in venous return from the head and upper extremities means the classic Glenn shunt or a bi-directional cavo-pulmonary anastomosis placed in childhood may fail to provide adequate palliation in the older patient.
 - pulmonary arteriovenous malformations (AVMs)—see below. AVMs result in a right to left shunt.

Pulmonary AVMs

The prevalence of pulmonary AVMs increases with time. By 10 years it is 10% in patients after a classic Glenn shunt. The prevalence as detected by pulmonary angiography rises to 20-25% over time, although the prevalence as detected by saline contrast injection into the pulmonary arteries may reach 71%. The median time after the cavo-pulmonary anastomosis to the development of pulmonary AVMs is 3.5 years.

The cause of pulmonary AVMs in patients with a cavo-pulmonary anastomosis is likely the absence of perfusion of the pulmonary vasculature by hepatic venous return. Similar pulmonary AVMs, with diffuse dilation of pre-capillary vessels, occur with cirrhotic liver disease. Supporting the concept that normal hepatic venous return bathing the pulmonary vasculature is protective against pulmonary AVMs is the resolution of pulmonary AVMs after a cavo-pulmonary anastomosis if the anastomosis is taken down and the SVC is reconnected to the right atrium, or if orthotopic heart transplantation is performed. (Pulmonary AVMs in cirrhotic liver disease reverses after liver transplant.)