

Mechanical Circulatory Support in Children with Acute Fulminant Myocarditis

Pranya Sakiyalak, M.D.*, Jarupim Soongswang, M.D.***, Somchai Sriyoscharti, M.D.*, Pansak Laksanabunsong, M.D.*

*Department of Surgery, **Department of Pediatrics, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok 10700, Thailand.

Siriraj Med J 2008;60:145-147

E-journal: <http://www.sirirajmedj.com>

Myocarditis is defined as inflammation of the myocardium accompanied by myocellular necrosis. Acute myocarditis must be considered in patients who present with a recent onset of cardiac failure or arrhythmia. Often there is a history of an antecedent flu-like illness.¹ Fulminant myocarditis is a distinct entity characterized by sudden onset of severe congestive heart failure or cardiogenic shock, usually following a flu-like illness. McCarthy et al. have shown that patients with fulminant myocarditis have a clinical course that is distinct from that of patients with acute myocarditis. Fulminant myocarditis is characterized by critical illness at presentation but excellent long-term survival. In contrast, patients with acute myocarditis are less ill initially but have a progressive course that leads to death or the need for cardiac transplantation. Having fulminant myocarditis was an independent predictor of transplantation-free survival even after adjustments were made for the severity of inflammation, age, and clinically relevant hemodynamic variables.²

Patients with fulminant myocarditis are critically ill at presentation. They can have a rapidly progressive course quickly resulting in death due to profound ventricular dysfunction. The mainstay of therapy for this form of myocarditis is supportive management. However, patients with rapidly progressing severe heart failure and shock may benefit from mechanical circulatory support. The most important factor leading to the decision to institute mechanical circulatory support is the observation that medical treatment is failing. Increasing inotropic requirements, accompanied by evidence of inadequate cardiac output such as poor cutaneous perfusion, oliguria, and systemic acidosis are unlikely to be reversed by further medical management in this condition. The need for excessive inotropic doses accompanied by significant ventricular ectopy is a particularly dangerous combination for which mechanical circulatory support should be strongly considered. Cardiac arrest, even of brief duration, is an ominous sign indicating that circulatory support should be considered. Overall, an aggressive stance toward the institution of mechanical circulatory support should be taken in these patients because of their tendency toward unpredictable and rapid deterioration.

Type of mechanical circulatory support

The options currently available for mechanical circulatory support are intra-aortic balloon pump (IABP), extracorporeal membrane oxygenation (ECMO), and ventricular assist device (VAD).

Intra-aortic balloon pump (IABP)

IABP is a non-flow generating counter pulsation type of assist. Intra-aortic balloon pump deflation in the systole leads to reduced ventricular after-load and reduced ventricular wall stress, thereby improving cardiac performance. Inflation of the balloon during the diastole increases the aortic diastolic pressure and hence coronary perfusion (Fig 1). An appreciable measure of the patient's own cardiac function must be present and the IABP augments this improving cardiac output to the tune of 15–20%. Therefore IABP is a poor form of support for total replacement of the function of a ventricle. It is extensively used in adults where it is a good form of support for coronary flow augmentation. Unfortunately, application of IABP in children has not been widespread due in part to difficulties with timing of counter pulsation in young patients, size of commercially available devices, and technical difficulty in

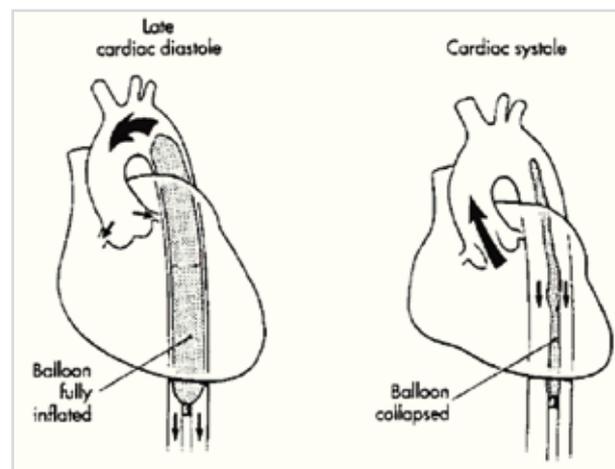


Fig 1. The intra-aortic balloon deflates in systole and inflates during diastole.

device insertion in small children. The aorta in children is also thought to have greater elasticity than that in adults. Hence, much of the energy generated during balloon filling may be transferred to the expansile aorta and diastolic augmentation may be dampened.³

Extracorporeal membrane oxygenation (ECMO)

Extracorporeal membrane oxygenation (ECMO) circulates and ventilates blood and can temporarily substitute for the entire circulation. However, like the heart/lung machine, ECMO activates blood elements to cause coagulation and inflammatory problems associated with cardiopulmonary bypass. A typical ECMO circuit is depicted in Fig 2. Deoxygenated blood is drained from the right atrium through a cannula inserted into the femoral or internal jugular vein and the blood that has been oxygenated extracorporeally is pumped back into the femoral artery, allowing total cardiopulmonary support.³ Thus, the heart and lung can be bypassed and hence rested, allowing time for the heart to heal in an environment of normal systemic perfusion and gas exchange. It is this flexibility of the ECMO circuit that makes ECMO versatile in supporting children with acute cardiac failure. Though results of ECMO have been excellent in selected patients, prolonged support is associated with significant morbidity, that is, peripheral vascular complications, hemolysis, and a bedridden state. In addition, ECMO does not decompress the left ventricle (LV) and is associated with increased LV wall stress. ECMO should be used for patients who present with circulatory collapse, cardiac arrest, or severe pulmonary failure with inability to oxygenate. In these circumstances, ECMO will allow stabilization of the patient with initial neurological, renal, and pulmonary recovery. If the heart shows signs of recovery over a few days, weaning to recovery is possible. If there is no improvement in cardiac function, the patient should be bridged, in general, to a ventricular assist device.

Ventricular Assist Device (VAD)

Ventricular assist devices are mechanical pumps that can perform the function of either one or both of the ventricles, restoring perfusion and maintaining end-organ function, while the heart recovers from the precipitating insult. Ventricular assist devices may be required for left ventricular support (LVAD), right ventricular support (RVAD) or for biventricular support (BIVAD).⁴ Pumps used in VAD can be divided into two main categories - continuous flow pumps and pulsa-

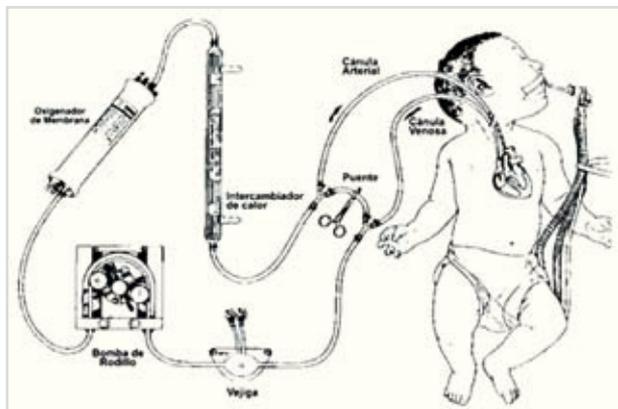


Fig 2. Depiction of a typical veno-arterial ECMO circuit.

tile pumps which mimic the natural pulsing action of the heart.

Continuous Flow Pumps

Continuous flow VADs normally use either centrifugal pumps or axial flow pumps. Centrifugal pumps and extracorporeal circuits have been used in infants and children since the development of a pediatric centrifugal pump head, Biomedicus (Medtronic, Eden Prairie, MN), in the late 1980s. Application of these systems is, however, limited to several days or a few weeks at most, and the patients must remain in the intensive-care unit. Centrifugal pumps are easy to use and inexpensive. The currently available centrifugal pump VADs in Thailand are BioMedicus Biopump, (Medtronic, Minneapolis, MN), Sarns centrifugal pump (3-M Health Care, Ann Arbor, MI, USA), and CentriMag[®] (Levitronix, Zürich, Switzerland). Based on vortex technology, these VAD systems use turbine spins of 10,000-20,000 rpm to create a flow of 5-6 l/min and have generally been applied for temporary assistance of stunned myocardium of the left ventricle (Fig 3). The centrifugal pumps are currently used for a short-term mechanical cardiac assist. Centrifugal pumps are commonly used in children with acute fulminant myocarditis with cardiogenic shock as a bridge to recovery. At our institute, we had 2 patients with acute fulminant myocarditis who were supported with centrifugal pumps and completely recovered of their cardiac function.

Axial pumps generally consist of electromagnetically coupled, integrated motor-impeller pump assemblies. An axial pump is a small pump that can be used for intermediate and long term circulatory support. The currently available pumps are DeBakeyVAD[®], INCOR[®], and Heartmate II. The more durable centrifugal pumps are also available for long term support such as Ventr-Assist[®] and DuraHeart[®].

Pulsatile Ventricular Assist Devices

All pulsatile pumps are basically membrane pumps

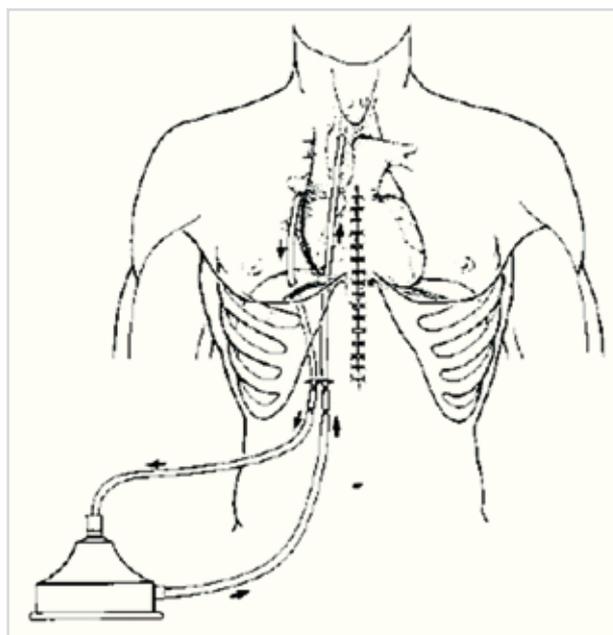


Fig 3. LVAD assistance with a Biomedicus centrifugal pump with cannulation of the left atrium and ascending aorta.

in which the membrane is moved by air, liquid or by a pusher-plate. The driving source (air, water or electricity) reaches the pump via a tube through the skin. The most often used indication of these devices is bridging to transplantation. Pulsatile devices can be located at the bedside, paracorporeally, or may be implantable. These devices, however, can only be used in adolescents with adequate body surface area (BSA) typically $>1.5 \text{ m}^2$ for implantable, and at least $> 0.7 \text{ m}^2$ for paracorporeal devices.³ Pulsatile pumps generate a pulsatile flow which reduces sympathetic nerve activity and peripheral vascular resistance, thus improving the microcirculation as well as organ function. In general, pulsatile VADs designed primarily for long-term bridge or destination therapy should be avoided if recovery within several weeks is expected.⁵ Such devices are more expensive, and more difficult to explant. The only pulsatile VAD currently available in Thailand is Thoratec paracorporeal ventricular assist device. Thoratec VAD can be used as LVAD, RVAD or BIVAD (Fig 4). It can be used for intermediate and long term circulatory support.⁶

In summary, children with acute fulminant myocarditis have an excellent long term prognosis. Full cardiac recovery is possible and even likely despite fulminant and catastrophic presentation. Paradoxically, patients who present as less ill have the worst long-term prognosis. Because of the reversibility of the ventricular dysfunction observed during the course of this disease in some patients and the option of cardiac transplantation for those whose ventricular function does not recover, aggressive measures to support these patients during the acute phase of their illness are justified. Providing temporary mechanical circulatory support until native ventricular function returns may be a successful strategy in the majority of these patients. Nevertheless, a subset of these patients will have refractory myocardial dysfunction requiring mechanical circulatory support. Pulsatile paracorporeal VADs have been validated as an effective strategy to keep children alive while awaiting heart transplantation.

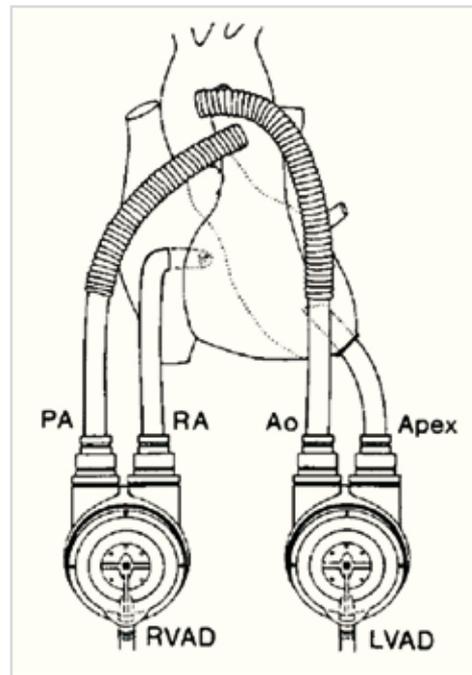


Fig 4. Biventricular support with Thoratec VADs

REFERENCES

1. Batra AS, Lewis AT. Acute myocarditis. *Curr Opin Pediatr* 2001;13:234-9.
2. McCarthy RE 3rd, Boehmer JP, Hruban RH, Hutchins GM, Kasper EK, Hare JM, Baughman KL. Long-term outcome of fulminant myocarditis as compared with acute (nonfulminant) myocarditis. *N Engl J Med* 2000;342:690-5.
3. Collision SP, Dagar KS. Mechanical circulatory support for acute cardiac failure in children: the options available. *Indian Heart J* 2006;58:345-9.
4. Duncan BW, Bohn DJ, Atz AM, French JW, Laussen PC, Wessel DL. Mechanical circulatory support for the treatment of children with acute fulminant myocarditis. *J Thorac Cardiovasc Surg* 2001;122:440-8.
5. Acker MA. Mechanical circulatory support for patients with acute-fulminant myocarditis. *Ann Thorac Surg* 2001;71:S73-6.
6. Sharma MS, Webber SA, Morell VO, et al. Ventricular assist device support in children and adolescents as a bridge to heart transplantation. *Ann Thorac Surg* 2006;82:926-33.