

Patient after Fontan Procedure Undergoing Scoliosis Surgery

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STEM CASE

A 9-year-old girl is scheduled for posterior spinal instrumentation for scoliosis with a rapidly progressive curve now measuring 55 degrees. The patient has had three surgeries for congenital heart disease, the last being a Fontan.

OBJECTIVES

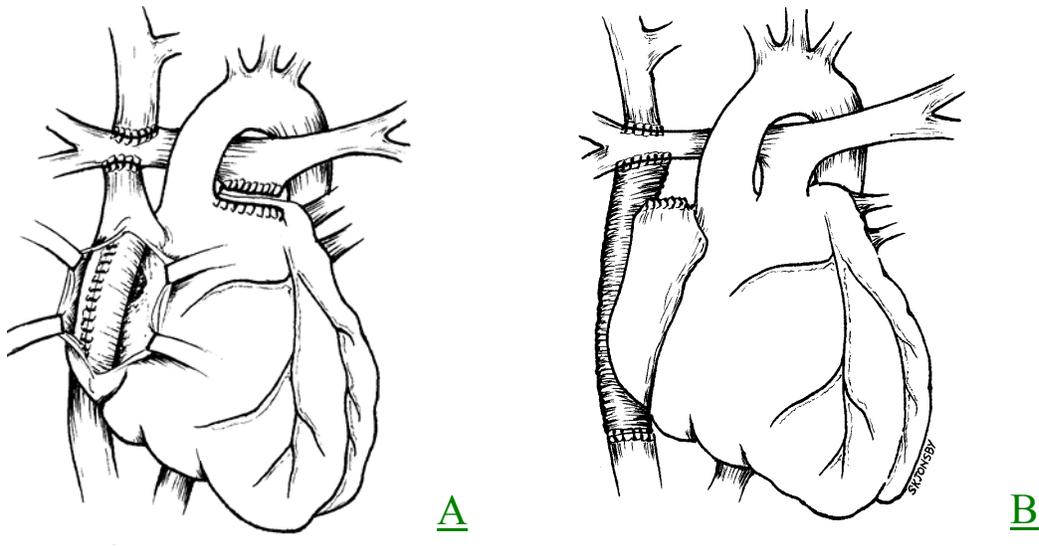
After case discussion participants should:

1. Understand cardiac anatomy and the physiology after the Fontan procedure.
2. Understand the long-term complications and prognosis after the Fontan procedure.
3. Develop an anesthetic plan for the induction and maintenance of anesthesia in patients after the Fontan procedure.
4. Understand the basic neurophysiologic principles of Somatosensory Evoked Potential (SSEP) monitoring.
5. Understand the pathophysiology of scoliosis and its effects on SSEPs.
6. Formulate an anesthetic plan for a patient undergoing scoliosis surgery with SSEP monitoring.
7. Understand the effects of anesthetics, physiologic changes, and neurologic injury on SSEPs.

Develop strategies to trouble-shoot and attempt to reverse adverse SSEP amplitude and latency changes that may occur during routine scoliosis operation

Problem Based Learning Discussion Model Discussion Outline:

1. What type of cardiac anatomy is treated with the Fontan procedure as final correction?



The figures show the most common modifications of the Fontan procedure. Figure A depicts a lateral tunnel Fontan in which blood from the inferior vena cava is routed to the pulmonary arteries through the native atrium, and the superior vena cava is directly connected to the right pulmonary artery. Figure B depicts an extracardiac Fontan in which blood from the inferior vena cava is routed to the pulmonary arteries through a conduit that is external to the heart, and the superior vena cava to pulmonary artery connection is the same.

2. What types of cardiac surgical procedures are typically performed prior to the Fontan procedure?

Initial Surgical Strategy for Single Ventricle Patients

Anatomy	Surgical Intervention
2 Semilunar Valves of adequate size, normal Aortic Arch	Pulmonary Artery Band
1 Semilunar Valve, normal Aortic Arch	BT Shunt
1 Semilunar Valve, hypoplastic Aortic Arch	Aortic Arch reconstruction with BT shunt, or Norwood Procedure
2 Semilunar Valves, aortic stenosis	Damus, Kaye, Stanzel with BT shunt (possible aortic arch reconstruction) or Palliative arterial switch
2 Semilunar Valves with pulmonary stenosis	No initial intervention required

Types of Single Ventricle Lesions

Single Left Ventricle
Tricuspid Valve Atresia
Double Inlet Left Ventricle
Unbalanced Complete Atrioventricular Canal (Hypoplastic right ventricle)
Heterotaxy Syndrome with right ventricular hypoplasia
Pulmonary Atresia with Intact Ventricular Septum

Single Right Ventricle
Hypoplastic Left Heart Syndrome and variants
Double Outlet Right Ventricle
Unbalanced Complete Atrioventricular Canal (Hypoplastic left ventricle)
Heterotaxy Syndrome with left ventricular hypoplasia

- 3. Explain general Fontan anatomy and physiology. What physiologic conditions are important to maintain?*
- 4. Various modifications of the Fontan procedure have been developed. What differences in pathophysiology can be anticipated from Fontan modifications?*
- 5. What is the significance of a fenestrated Fontan? Would you recommend closure of the fenestration prior to scoliosis surgery?*
- 6. What hemodynamic problems can be anticipated during anesthesia and how will they affect your management?*
- 7. Is it necessary to give antibiotic prophylaxis?*

8. How will you monitor the patient's hemodynamic status?

9. Should external defibrillator/cardioversion/pacing pads be applied prior to the induction of anesthesia? Where should they be applied?

10. What are the basic principles of Somatosensory Evoked Potential Monitoring?

somatosensory-evoked potentials (SSEP)	Responses to electrical stimulation of a sensory peripheral nerve
motor-evoked potentials (MEP)	Measurement of compound muscle action potentials following direct stimulation of a descending motor nerve
brainstem auditory evoked potentials (BAEP)	brainstem recording in response to auditory stimuli
visual-evoked potentials (VEP)	Occipital recordings of visual flashing monocular stimuli
Transcranial motor evoked potential (TcMEP)	Recordings of the contralateral motor groups after transcranial stimulation of the motor cortex

11. What other options for spinal cord monitoring are available?

12. What are the advantages and limitations of SSEP monitoring in comparison to other types of monitoring?

13. What are the different classifications of scoliosis, and how might they affect baseline SSEP recordings?

Congenital	Vertebral and/or rib anomalies and spinal dysraphism
Idiopathic-Infantile	<3 years
Idiopathic Juvenile	3-10 years
Idiopathic-Adolescent	10 years
Neuromuscular Disease	Cerebral palsy, poliomyelitis, myopathies, syringomyelia, Friedrich's ataxia etc.
Traumatic	Fractures, radiation, burns, surgery
Syndromes	Neurofibromatosis, Marfan's, osteogenesis imperfecta, mucopolysaccharidosis, rheumatoid arthritis
Neoplastic	

14. What is the justification for performing SSEP monitoring during scoliosis surgery

15. How well does SSEP monitoring detect an injury that may cause paralysis?

16. What are the effects of the different anesthetics on SSEPs?

You give a fluid bolus to the child and proceed with intravenous induction of anesthesia. After successful intubation, the patient is hypotensive with a blood pressure 70/40 mmHg and heart rate 120 bpm.

17. What is your differential diagnosis?

Following CVP guided fluid administration, the patient remains hypotensive and the oxygen saturation has decrease to 91% with inspired FiO₂ at 100 %.

18. What is your differential diagnosis? Would a Valsalva maneuver help? What problems specific for Fontan physiology can cause this hypotension and hypoxia?

19. What can be done to resolve the problem?

The child's starting hematocrit is 49%. You want to minimize blood exposure.

21. What methods are available to decrease bleeding? Does the risk/benefit of these maneuvers change in the Fontan patient?

SSEP monitors are placed, bilateral baseline SSEP waveforms are collected, and the latencies and amplitudes are recorded.

22. Where are SSEP stimulating and recording electrodes placed?

23. How are SSEP waveforms generated?

24. What SSEP waveforms are recorded?

Soon after skin incision, there is a sudden change in the recordings.

25. What are the possible causes for these sudden changes, and how could you correct them?

26. What is considered a significant change in latency and in amplitude?

The surgeon now performs the instrumentation, and while placing a laminar wire there is again a SSEP change.

27. Now what do you suspect potential causes may be?

The procedure is near completion and the nurse requests to know if a ventilator should be prepared for the patient.

28. What are advantages and disadvantages of providing post-operative mechanical ventilatory support? What parameters would affect your decision?

29. What post operative pain management would you plan? What are advantages and disadvantages of different methods?

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