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European Journal of Cardio-thoracic Surgery 20 (2001) 114–119

EUROPEAN JOURNAL OF CARDIO-THORACIC SURGERY

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The enhancement of hemodynamic performance in Fontan circulation using pain free spontaneous ventilation ☆

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Received 8 October 2000; received in revised form 28 March 2001; accepted 6 April 2001

Abstract

Objective: Positive pressure ventilation is known to have a deleterious effect on pulmonary blood flow in patients with Fontan physiology. We evaluated the hemodynamic effects of pain free, spontaneous, non-positive pressure ventilation in patients undergoing Fontan staging procedures or completion. Fontan procedures, with creation of low pressure passive pulmonary circulation. **Methods**: Between May 1997 and May 1999 50 consecutive patients undergoing either bi-directional Glenn (BDG, $n = 23$) or completion Fontan ($n = 27$), were managed with early extubation. Anaesthetic management included continuous narcotics, caudal block, epidural block, or hyperbaric spinal. Post-operative management included low dose dopamine (3 mcg/kg per min), nitro-glycerine (0.3 mcg/kg per min) and nitroprusside (0.3 mcg/kg per min). Post-operative management was identical for all patients. Twelve patients were randomly selected to undergo continuous cardiac output and cardiac index (CI) determinations utilizing extra vascular Doppler probes placed on the ascending aorta, allowing for continuous aortic diameter and Doppler wave form velocity recordings. All patients were extubated either in the operating room or within one hour post-operatively. There were no deaths and no complications in the series. Mean length of stay (LOS) for BDG was 4.3 ± 0.5 days. Mean LOS for Fontan patients was 11 ± 4 days. **Results**: Mean pulmonary artery pressure (MPAP) fell from 19 ± 3.464 pre-extubation to 14 ± 3.271 immediately post-extubation, 13.2 ± 2.261 6 h post-extubation, and 11.7 ± 2.146 12 h post-extubation. All decreases in MPAP post-extubation were significant ($P = < 0.05$). CI pre-extubation was 3.25 ± 1.09, immediately post-extubation 5.05 ± 1.297, 12 h post-extubation 6.225 ± 1.19. All increases in CI post-extubation were significant ($P = <0.05$). **Conclusion**: Resumption of pain free, spontaneous, non-positive pressure ventilation enhances hemodynamic performance in patients with Fontan circulation and clearly improves outcome.

Keywords: Bi-directional Glenn shunt; Fontan

1. Introduction

The perioperative and post-operative management of patients undergoing creation of bi-directional Glenn shunts or completion Fontan procedures can be problematic and controversial. Management of these patients is largely dependent upon an individual institutions philosophy with regard to hemodynamic and ventilation parameter manipulation. With a passive pulmonary circulatory arrangement, decreasing pulmonary vascular resistance is of paramount importance, and is very much dependent upon adequate ventilation, usually achieved through mechanical ventilation. This is offset by the potentially detrimental effects of endotracheal intubation and positive pressure ventilation.

For the past 11 years the author has practised a policy of extremely early extubation with resumption of spontaneous ventilation as quickly as possible at the completion of the Fontan or Fontan staging procedure. Patients were extubated before leaving the operating room or immediately upon arrival in the intensive care unit. The clinical impression existed that these patients simply did better hemodynamically than patients receiving positive pressure ventilation. There has been little data to support this clinical impression.

The purpose of this study was to retrospectively review all patients undergoing Fontan or Fontan staging procedures over a 24-month period beginning in May of 1997, and to determine how a protocol and policy of early extubation effects hemodynamic performance.

2. Materials and methods

2.1. Patient population

Fifty consecutive patients undergoing either bi-direc-

☆ Presented at the 14th Annual Meeting of the European Association for Cardio-thoracic Surgery, Frankfurt, Germany, October 7–11, 2000.

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1010-7940/01/$ - see front matter
PII: S1010-7940(01)00757-6

tional Glenn shunts or completion Fontan procedures between May of 1997 and May of 1999 were included in this study group. All patients presenting for and undergoing these procedures were included, and none were excluded. Twenty-three patients underwent creation of bi-directional Glenn shunts, while twenty-seven patients underwent completion Fontan procedures. Patients undergoing bi-directional Glenn shunts ranged in age from 3.5 to 18 months, with a mean and median age of 6–6.5 months. The weights of these patients ranged from 5.5 to 11.1 kg, with a mean ± standard deviation ($M \pm SD$) of 7.6 ± 1.231 kg, and a median of 7.4 kg. Patients undergoing Fontan procedures ranged in age from 18 to 60 months, with a mean age of 27.2 ± 5 months, and ranged in weight from 9.25 to 17.3 kg, with a mean weight of 14.49 ± 8.083 kg. Patient characteristics are summarized in Table 1.

2.2. Statistical analysis

Mean pulmonary artery pressures (MPAP) were recorded continuously and sampled at pre selected time intervals for all patients in this study. Cardiac index (CI) was recorded continuously from 12 randomly selected patients and sampled at preselected time intervals concomitantly with the mean pulmonary artery pressures. Repeated Measures Analysis of Variance (GraphPad Instat V2.02, GraphPad Software, Suite 203, 10855 Sorrento Valley Road, San Diego, CA 92121) was used to compare the means of the matched MPAP and CI groups. Student–Newman–Kuhls Multiple Comparisons Test was then utilized for additional analysis. All values are expressed as mean ± standard deviation. *P*-values are presented with one or two significant digits where possible and appropriate, with $P < 0.05$ considered significant.

2.3. Anaesthetic management

Intra-operative anaesthetic management consisted of continuous narcotics with fentanyl and versed, and caudal block, epidural block or hyperbaric spinal anaesthesia. Anaesthetic preference was determined entirely by the individual anaesthesiologist.

2.4. Operative management

All operations were performed using cardiopulmonary bypass. Three patients undergoing bi-directional Glenn shunts underwent deep hypothermia, because of the need to reconstruct the aortic arch in one, and the need to perform Damus–Kaye–Stansel anastomoses in two. The remaining twenty patients undergoing bi-directional Glenn shunts underwent normothermic cardiopulmonary bypass with selective cannulation of the superior vena cava and the right atrium. Three patients required enlargement of the atrial septal defect. In these patients, the inferior vena cava was cannulated at its junction with the right atrium to allow for the placement of Rummell tourniquets. While on normothermic bypass, and the caval tapes occluded, ventricular fibrillation was induced, the right atrium opened, the atrial septum excised, and the right atrium then closed. Ventricular fibrillation times did not exceed three minutes, after which the bi-directional Glenn anastomosis was then performed.

All 27 patients undergoing Fontan completion underwent normothermic cardiopulmonary bypass, with no aortic cross clamping, and no cardioplegia being administered. All patients received extracardiac conduits using tubular grafts of expanded polytetrafluroethylene ranging in size from 14 to 18 mm in diameter. Anastomoses were constructed between tube graft and the transected inferior vena cava at its junction with the right atrium (leaving a cuff of atrial tissue), and the pulmonary arterial confluence. Total operative times and cardiopulmonary bypass times for both groups of patients are tabulated in Table 2.

All patients in this series underwent modified ultrafiltration prior to decannulation. All patients were monitored using superior vena cava (CVP) lines and peripheral arterial lines placed percutaneously during anaesthesia induction. No left atrial lines were placed. In addition, twelve randomly selected patients from both the bi-directional Glenn shunt group and the completion Fontan group underwent placement of a prototype extravascular Doppler probe on the ascending aorta (Applied Biometrics, Burnsville, MN). The probes contained two ultrasound transducers, which allowed for continuous aortic diameter readings and continuous Doppler velocity readings. The wiring for the probes was brought out transcutaneously through the thoracic wall and interfaced to a screen, which allowed continuous display of blood velocity waveforms, cardiac output, and cardiac index. The probes were then extracted on the morning following surgery.

Table 1
Age in months and weight in kilograms of patients in both study groups

	Age (months)	Weight (kg)
BDG[a]	3.5–18	5.5–11.1
	6.5±1.5 ($M \pm SD$)	7.6±1.231 ($M \pm SD$)
Fontan	18–60	9.3–47.0
	27.2±0.5 ($M \pm SD$)	14.49±8.083 ($M \pm SD$)

[a] BDG, bi-directional Glenn shunt.

Table 2
Cardiopulmonary bypass times and total operative time in minutes for patients in both groups

	CPB (min)	Total operative time (min)
BDG[a]	35–140	106–262
	78.5±31.356 ($M \pm SD$)	175±45.04 ($M \pm SD$)
Fontan	42–200	147–370
	81.85±35.892 ($M \pm SD$)	207±57.6 ($M \pm SD$)

[a] BDG, bi-directional Glenn shunt.

2.5. Post-operative management

All patients were managed with early extubation, either in the operating room, or within 1 or 2 h following arrival in the intensive care unit. Patients receiving epidural block or hyperbaric spinal received no additional narcotics. The remaining patients received supplemental doses of fentanyl and versed intravenously as needed. All patients received by continuous intravenous infusion nitro-glycerine at 0.3 mcg/kg per min, sodium nitroprusside at 0.3 mcg/kg per min, and dopamine at 3 mcg/kg per min. The dopamine infusion remained constant at 3 mcg/kg/min for the first 24 h post-operatively. No patients required additional inotropic support either by increasing the level of dopamine infusion, or through the addition of other inotropic agents. Systemic systolic blood pressures were maintained at 85–110 mm of mercury in all patients through after-load reduction, by varying the infusion rates of nitroprusside and nitro-glycerine. All patients were monitored with continuous pulse oximetry, with oxygen saturations being maintained greater than 75% in the bi-directional Glenn patients, and greater than 95% in the completion Fontan patients. Fluid replacement for all patients was through crystalloid solutions administered at two-thirds of calculated maintenance based upon body surface area. No patients received diuretics during the first 24 h post-operatively. Post-operative bleeding was negligible in all patients, so no additional infusions of blood or blood products were required. In all patients avoidance of CO_2 retention and avoidance of pain and agitation were considered of paramount importance.

3. Results

Central venous pressure becomes the mean pulmonary arterial pressure in the presence of a direct cavo-pulmonary connection, and will henceforth be referred to as mean pulmonary arterial pressure. Mean pulmonary arterial pressure (MPAP) fell from 19 ± 3.464 pre-extubation to 14.83 ± 3.271 immediately following resumption of spontaneous respiration and extubation. This decrease was significant ($P < 0.001$). MPAP 6 h post-extubation was 13.25 ± 2.261, a decline that was also significant ($P < 0.001$) compared to pre-extubation. The decline in MPAP continued throughout the first 12 h, with MPAP 12 h post-extubation 11.67 ± 2.146, a decrease that was also significant ($P < 0.001$) compared to pre-extubation. Twelve hours following surgery the monitoring lines were removed. Cardiac index rose from 3.225 ± 1.09 pre-extubation to 5.075 ± 1.297 immediately following resumption of spontaneous respiration and extubation, an increase that was significant ($P < 0.05$). Cardiac index 12 h following extubation increased to 6.525 ± 1.19, an increase that was significant ($P < 0.01$) compared to pre-extubation, and significant ($P < 0.05$) compared to immediately post-extubation. The cardiac index probes were removed 12 h post-extubation. The hemodynamic findings are summarized in Table 3, Table 4, and Fig. 1.

There were no operative deaths in this series. All patients were discharged from the hospital. Two late deaths have occurred in the bi-directional Glenn group. One death was from progressive pulmonary venous obstructive disease in a patient with complex heterotaxy who had undergone correction of an interrupted arch and total anomalous venous connection as a new-born. The second death was in a patient with hypoplastic left heart syndrome that expired during an acute infection with respiratory syncytial virus. There have been no late deaths in the Fontan patients. Length of stay for the bi-directional Glenn shunt patients was 4–5 days, with a

Table 3
Mean pulmonary artery pressure (MPAP) measured immediately prior to extubation (pre), immediately following extubation (post), and at intervals post-extubation

Pre	Post	6 h post	12 h post
19.0 ± 3.464 ($M \pm$ SD)	14.8 ± 3.271 ($M \pm$ SD)	13.25 ± 2.261 ($M \pm$ SD)	11.67 ± 2.146 ($M \pm$ SD)
Comparison	Mean difference	Q	P value
12 h post vs. 6 h post	−1.583	2.472	ns, $P > 0.05$
12 h post vs. post	−3.167	4.943	$^{**}P < 0.01$
12 h post vs. pre	−7.333	11.447	$^{***}P < 0.001$
6 h post vs. post	−1.583	2.472	ns, $P > 0.05$
6 h post vs. pre	−5.750	8.976	$^{***}P < 0.001$
Post vs. pre	−4.167	6.504	$^{***}P < 0.001$
Difference	Mean difference	Lower 95% CI	Upper 95% CI
12 h post–6 h post	−1.583	−3.428	0.2610
12 h post–post	−3.167	−5.391	−0.9419
12 h post–pre	−7.333	−9.786	−4.880
6 h post–post	−1.583	−3.428	0.2610
6 h post–pre	−5.750	−7.975	−3.525
Post–pre	−4.167	−6.011	−2.322

Table 4
Cardiac index (CI) measured immediately prior to extubation (pre), immediately following extubation (post), and 12 h post-extubation

Pre	Post	12 h Post	
3.225 ± 1.090 ($M \pm$ SD)	5.075 ± 1.297 ($M \pm$ SD)	6.525 ± 1.190 ($M \pm$ SD)	
Comparison	Mean difference	Q	P value
Pre vs post	−1.850	4.678	$^{*}P < 0.05$
Pre vs 12 h post	−3.300	8.345	$^{**}P < 0.01$
Post vs 12 h post	−1.450	3.667	$^{*}P < 0.05$
Difference	Mean difference	Lower 95% CI	Upper 95% CI
Pre–post	−1.850	−3.219	−0.4813
Pre–12 h post	−3.300	−5.016	−1.584
Post–12 h post	−1.450	−2.819	−0.08131

mean of 4.3 days. Length of stay for the Fontan patients was 4–20 days, with a mean of 11 days.

4. Discussion

In 1954, Glenn and Patino described direct delivery of vena caval blood into the pulmonary arterial circulation through an azygos vein to pulmonary artery anastomosis [1]. This was followed in 1958 by a description of an anastomosis between the superior vena cava and distal pulmonary artery [2].

In 1966, Haller and Associates [3] performed experimental studies on laboratory animals in which an end to side anastomosis between a transected superior vena cava and the proximal right pulmonary artery was accomplished. This was not utilized clinically until some time later.

In 1971 Fontan and Baudet [4,5] described direct atrial pulmonary connection for the treatment of tricuspid atresia. Other authors too numerous to include have proposed modifications of the operation described by Fontan and Baudet in order to definitively palliate other forms of single ventricle.

In 1985, Hopkins and Associates [6] introduced clinically the concept of the bi-directional Glenn shunt which has proven to be an extremely important anatomic and physiologic adjunct in achieving a total caval pulmonary connection.

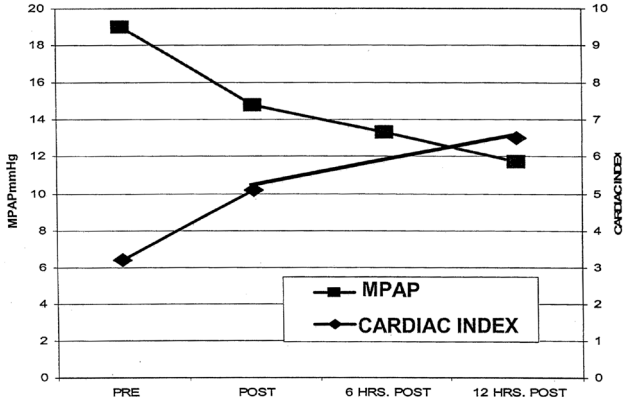

Fig. 1. MPAP plotted against CI immediately prior to extubation (pre), immediately following extubation (post), and at intervals post extubation.

Other authors have continued to investigate the hemodynamics of variable degrees of right heart bypass. Although these studies were performed in experimental animals, their clinical relevance is quite important [7].

While most studies and series of Fontan patients have dealt with patient selection and operative technique, few studies have focused specifically on perioperative and post-operative management of these patients. It has long been recognized that the post-operative management of a patient with passive pulmonary circulation may be problematic. In 1981, Heck and Doty even proposed phasic external lower body compression as an adjunct to assisted circulation, feeling that this technique diminished fluid sequestration and improved cardiac output [8].

Standard textbooks do not dwell extensively on post-operative management, but mention that mechanical ventilation may be required while hemodynamic and ventilatory parameters are manipulated. Extubation is usually accomplished within 6–48 h of operation [9].

Many institutions are reluctant to extubate patients after creation of a bi-directional Glenn shunt or after a Fontan operation out of concern that atalectasis and subsequent pulmonary compromise would impair hemodynamic performance. This would certainly seem to be true with a passive pulmonary circulatory arrangement.

Spontaneous respiration creates a negative intrathoracic pressure with inspiration, which increases systemic venous return. If spontaneous respiration can be achieved without pain and subsequent splinting, it is understandable that hemodynamics would be improved, especially if the pulmonary circulation is dependent upon systemic venous return.

Working in conjunction with anaesthesiologists for the past 11 years, the author has practised a policy of extremely early extubation in patients undergoing bi-directional Glenn shunts or Fontan procedures. It has been our standard of care to extubate patients either in the operating room, or immediately upon arrival in the intensive care unit, once anaesthetic agents were sufficiently metabolized. Through the

years we had noticed a sharp decline in central venous pressure or mean pulmonary artery pressure immediately post-extubation, and a steady decline in central venous pressure or mean pulmonary artery pressure during the first 12 h post-extubation. Since these patients all have a common atrium, any other form of invasive monitoring was unnecessary and care was simplified.

The availability of prototype cardiac output/cardiac index probes enabled further investigation and documentation of a clinical practice that was already well established. The probes and the software had been extensively investigated in laboratory animals. We would have preferred to do cardiac output/cardiac index determinations in more patients, but only 12 prototype probes were available. The probe is being reconfigured to allow for easier extraction, and it is not yet commercially available.

Use of the probe, coupled with mean pulmonary artery pressure measurements, did enable us to demonstrate that pain free spontaneous respiration does indeed enhance hemodynamic performance in patients with a passive pulmonary circulatory arrangement.

We would conclude that perioperative anaesthetic and post-operative analgesic management plays an important role in the overall management of patients with passive pulmonary circulation. Resumption of pain free spontaneous respiration does enhance hemodynamic performance in these patients. We would also infer that, while conduct of operation remains of paramount importance, operative technique, intraoperative anaesthetic, and post-operative analgesic management all interact to contribute to overall results and outcomes.

Acknowledgements

The authors would like to acknowledge the contributions of Gary Grist, R.N., for The Children's Mercy Hospital data base and statistical preparation, and Arta Williams, for preparation of the manuscript.

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Appendix A. Conference discussion

Dr R. Neirotti *(Grand Rapids, MI, USA)*: Having had a longstanding interest in early extubation, I very much enjoyed your presentation. We have reviewed recently a series of 850 patients and the overall extubation rate was 80%. I agree with you that those patients with single ventricle physiology really benefit from extubating them in the operating room or upon arrival in the intensive care. Now, if you use intraoperative transesophageal echocardiography, you would see changes in the flow pattern that will explain the hemodynamic findings that you have shown; that is, the decrease in the CVP and the increase in the cardiac output.

Dr Lofland: We have not utilized TEE in this particular patient group, but it's fascinating to think that we could have that kind of documentation of increased flow with creation of a negative intrathoracic pressure.

Dr B. Maruszewski *(Warsaw, Poland)*: I would like to comment that recently, even if we find in some patients, complex patients, an impaired cardiac output immediately after surgery, we still will treat them using very early extubation protocol, because we believe and have experienced that it helps in early post-op course.

I have a question which is not directly related to the aim of the study, but you mentioned that you have put external conduits in Fontans between 14 mm and 18 mm in size. Do you have any evidence, or how long do you think it is going to last, specifically 14 mm? We wouldn't put any below 18 really, because then we would expect very early redo.

Dr W. Brawn *(Birmingham, UK)*: I must say that I was wondering about the size of those conduits as well.

Dr Lofland: Many of our patients are done at a fairly young age and are fairly small patients.The 14 mm size seemed to be an appropriate size to not impede flow from the cava into the pulmonary circulation and not act as an additional capacitance circuit with stasis of blood, but we have no evidence to support the size use. We have not redone any of these procedures. And I want the flow through the conduit to be very rapid.

Dr Maruszewski: I think if you have a patient in whom you necessarily have to put conduit of 14 mm in size, we would consider this patient either for staged or for an intracardiac Fontan type of operation.

Dr F. Lacour-Gayet *(LePlessis Robinson, France)*: I presume from your presentation that you were able to measure the cardiac output. I think this is very interesting in post-operative management. Could you comment a little bit on the technique that you are using and how you eventually take out the probe that you have to put in?

Dr Lofland: Yes. The cardiac output and index probes that we used are not commercially available, and all 24 probes that we had were prototypes. We're very satisfied with the algorithms that are utilized for calculation of cardiac output and index, both in animal models and in the human arena. While being satisfied with the algorithms, we have withdrawn the probe to make the implantation and extrication a bit less intimidating. Basically the probe is sutured to the ascending aorta using a wiring technique that allows for withdrawal of the wires while leaving the sutures in place, so the probe essentially separates from the wires and then is brought out through a little transcutaneous incision that is roughly the size of a 14 French chest tube. It's just pulled out transcutaneously. But the probes that we used will bear very little resemblance to the ones that will be commercially available. They will be more streamlined.

Dr Brawn: I would like to ask one more thing relating not necessarily to

your paper, but you commented that the length of hospital stay has been decreasing over time. Is that just with experience, or have you introduced any other particular measures to bring about that in terms of reducing incidence of effusions or length of time effusions can occur and so forth?

Dr Lofland: We haven't done anything to change our operative technique, conduct of operation, or anaesthetic management. We have made no alterations whatsoever in our post-operative management except to introduce a different pleural drainage system which interfaces to a suction bulb. We don't put any of these patients on waterseal drainage systems anymore. We have a very extensive patient education apparatus in place, so patients can be discharged early, leaving the drainage system in place, and actually return to clinic to have the drainage system removed in the clinic.

Dr A. Corno *(Lausanne, Switzerland)*: 24 h after surgery you have 11 mmHg as the average pressure in the vena cava system. That means you have a perfect indication, perfect operation, and good ventricular function. Was this a series of consecutive patients or not? In this same period did you turn out any patient for cavopulmonary connection because of a borderline indication, poor ventricular function or so forth?

Dr Lofland: No. These were 50 consecutive patients. These were all patients undergoing this procedure. No patients were excluded from the study. These were all the patients who had those procedures performed at our institution during that time period.