Fontan “Ten Commandments” Revisited and Revised

Herbert J. Stern

Received: 12 July 2010 / Accepted: 17 September 2010 / Published online: 20 October 2010
© Springer Science+Business Media, LLC 2010

Abstract Choussat’s “Ten Commandments,” which describes the components of an ideal Fontan candidate, was first published in 1977. Despite the wisdom in these commandments, it is clear from a historic perspective that total compliance with all criteria does not necessarily portend excellent long-term survival. I believe the end point of the original commandments should be modified to include improvement in long-term survival. I suggest the following single commandment: “Thou Shalt Be Perfect.”

Keywords Fontan · Single Ventricle · Aorto-Pulmonary Collaterals · Resynchronization Therapy · Arch Obstruction · Coil Occlusion · Endovascular Stenting

Choussat’s “Ten Commandments,” which describes the components of an ideal Fontan candidate, was first published in 1977 [7, 8] (Table 1). These guidelines, modified slightly by various centers, have served clinicians well these past 33 years by helping to determine which patients could safely be staged toward Fontan palliation with a high probability of success. Despite the wisdom in these commandments, it is clear from a historic perspective that total compliance with all criteria does not necessarily portend excellent long-term survival because Kaplan-Meier survival curves demonstrate a disturbing attrition trend [10]. Given the advancements made in catheter/interventional techniques and in surgical and hybrid techniques, as well as advancements in imaging modalities to guide invasive techniques and newer pacing technologies, I believe that

the end point of the original commandments should be modified to include improvement in long-term survival. I suggest the following single commandment: “Thou Shalt Be Perfect.”

At the recent World Congress Pediatric Cardiology meeting in Cairns, Australia, Dr. James Wilkinson presented a lecture entitled, “The Fontan Ten Commandments: Have They Doubled or Halved?” His conclusion was that the original 10 commandments could be condensed to four.

He argued that age <4 years is now the norm because earlier palliation has become standard in most centers. Abnormalities of systemic venous return can be overcome with bilateral cavopulmonary shunts and rerouting surgical procedures. Right atrial volume is not important because the classic Fontan has been replaced with either the lateral tunnel or extracardiac baffle. Small and/or distorted pulmonary arteries can be plastied from hilum to hilum, and regurgitant mitral valves can be repaired at the time of surgery (the original criteria were applied to patients with tricuspid atresia only).

Even Dr. Wilkinson’s modifications, however, do not take into account subtle residual defects that can conspire to induce irreversible changes to the myocardium, thus leading to attrition [3, 15, 20, 29, 40]. Without highlighting these residual defects, clinicians, when contemplating repair, often apply the same criteria to univentricular hearts that they would apply to biventricular hearts, which obviously have more reserve. Common examples include mild aortic arch obstruction, branch pulmonary stenosis, mild to moderate degrees of atrioventricular and semilunar valve insufficiency, aortopulmonary collateral flow, and abnormalities of rhythm with or without dyssynchronous ventricular contraction.

Mild degrees of aortic arch obstruction are common after Norwood palliation for hypoplastic left heart

H. J. Stern (✉)
Stern Cardiology, Quail View Road, Charlotte, NC, USA
e-mail: tocathu@gmail.com

Table 1 Choussat's ten commandments

Choussat et al. (1977) delineated selection criteria to define an ideal candidate for a Fontan procedure. They described the 10 following criteria, which are occasionally and facetiously referred to as the Ten Commandments for an ideal Fontan operation result.

Age >4 years
Sinus rhythm
Normal systemic venous return
Normal right atrial volume
Mean pulmonary artery pressure <15 mm Hg
Pulmonary arteriolar resistance <4 Wood units/m ²
Pulmonary artery–aorta ratio >0.75
Left-ventricular ejection fraction >0.60
Competent mitral valve
Absence of pulmonary artery distortion

syndrome [5]. The generally accepted criterion for intervention in a biventricular heart is a peak-to-peak gradient of 20 mm Hg [3, 12, 30, 44]. This criterion, although arbitrary by itself, has been even more inappropriately applied to univentricular hearts, despite the fact that studies have shown that peak systolic gradients <20 mm Hg can cause significant diastolic dysfunction [23, 41]. With advances in catheter/interventional techniques alone, or with hybrid (surgical/catheter) techniques, it is possible to completely abolish aortic arch gradients. Holzer et al. demonstrated that even complex transverse aortic arch obstructions can be successfully approached with open cell–design stents, thus allowing crossed head vessels to be redilated if needed [13]. Stern et al. demonstrated the safety and efficacy of a new premounted stent (which can be redilated to 20 mm) in dilating vascular stenosis in infants and toddlers [37]. Is it reasonable, therefore, to accept any arch gradient in a univentricular heart, which is already exposed to increased afterload [32, 38]?

Pulmonary artery narrowing or distortion can be approached in the same manner as aortic arch obstructions, equalizing pulmonary blood flow and decreasing already elevated caval and lymphatic pressures, in addition to reducing the total resistance the single ventricle faces [28, 45].

With the advent and common application of three-dimensional (3-D) echocardiography (echo) and magnetic resonance imaging, it is clear that better imaging techniques provide the surgeon with a better understanding of atrioventricular and semilunar valve anatomy and, in turn, the pathology behind regurgitation, which is unique to each patient [1]. This allows the surgeon, as never before, to plan and execute surgery to either eliminate or make trivial these valvular abnormalities [27, 31]. We should not accept more than a mild degree of regurgitation because volume

loading will result in accelerated failure of the Fontan circulation [22].

Aortopulmonary collateral vessels are known to be detrimental in single-ventricle patients, although their anatomy is poorly described in the literature, and the technique to obliterate these vessels has only recently been published [2, 14, 18, 35, 36]. I believe elimination of chronic volume loading and competitive flow, along with cavopulmonary shunting ideally by age 4 months, is absolutely essential [36]. In combination with managing the residual defects previously noted, this should allow for better and longer-lasting palliation.

Abnormalities of heart rhythm and rate, including re-entrant atrial arrhythmias, bradyarrhythmias, and chronotropic incompetence, are extremely common in post-Fontan patients [9]. Fontan patients today will generally have either a lateral tunnel or extracardiac baffle with or without fenestration. The common practice of transcatheter closure of fenestrations in those that have not spontaneously closed makes the transvenous route for pacemaker leads problematic at best for most patients. The combination of atrial arrhythmias and underlying sinus and/or junctional bradycardia makes therapy with antiarrhythmic medications extremely challenging. Surgeons should plan for this eventuality by placing steroid-eluting epicardial leads at the time of the Fontan repair [39].

In addition, resynchronization therapy using multisite pacing has been shown to be effective in univentricular hearts with prolonged QRS durations and dysynchronous ventricular contraction [11, 17, 19, 33, 34]. Although it may be impossible to predict the manner of dysynchrony at Fontan surgery, which would require epicardial leads later, we are aware of the development of an epicardial pacemaker multilead array incorporated into a cardiac harness, which could be placed at Fontan surgery and used, when appropriate, at a later date [8]. I agree with Cecchin et al. that more sensitive modalities beyond ejection fraction and QRS duration must be used when assessing the efficacy of this technology [4]. Promising alternatives include tissue Doppler imaging, tissue synchronization imaging, 3-D echo, and isovolumic acceleration times [4, 42]. By anticipating these problems in a proactive manner at Fontan surgery, we could avoid months and years of pathologic functioning resulting in neurohormonal activation, apoptosis, and the eventual downward spiral of the health of these patients [6, 21].

Finally, optimal medical management of these complex patients is only just receiving the prospective randomized trial attention it deserves. Thus, management varies widely from one center to the next, with practitioners relying on their own experience, anecdotal reports, and underpowered randomized trials. Can heart failure algorithms in adults without congenital heart disease be successfully

extrapolated to this cohort of patients [24, 43]? Will phosphodiesterase-5 inhibitor drugs, such as sildenafil, demonstrate efficacy not only as a vasodilator but a primary inotropic drug as well [25]? And what is the most prudent regimen to keep these patients from developing thrombi [16, 26]? Only by centers pooling their patients into large, prospective, randomized trials will we achieve the diagnostic power to answer these critical questions.

Patients with single-ventricle anatomy present a unique challenge to the cardiologist caring for them. Only by pre-emptive and aggressive management will clinicians begin to see a positive impact on Kaplan-Meier survival curves. I recommend that Choussat's "Ten Commandments" be revised to one, simple commandment: "Thou Shalt Be Perfect." We have the necessary technology, and even if we cannot be perfect, we can strive to be.

References

1. Apostolakis EE, Baikoussis NG (2009) Methods of estimation of mitral valve regurgitation for the cardiac surgeon. *J Cardiothorac Surg* 4:34
2. Ascuitto RJ, Ross-Ascuitto NT (2004) Systemic-to-pulmonary collaterals: a source of flow energy loss in Fontan physiology. *Pediatr Cardiol* 25(5):472–481
3. Beekman RH, Rocchini AP, Behrendt DM, Rosenthal A (1981) Reoperation for coarctation of the aorta. *Am J Cardiol* 48:1108–1114
4. Cecchin F, Frangini PA, Brown DW, Fynn-Thompson F, Alexander ME, Triedman JK et al (2009) Cardiac resynchronization therapy (and multisite pacing) in pediatrics and congenital heart disease: five years experience in a single institution. *J Cardiovasc Electrophysiol* 20:58–65
5. Chessa M, Dindar A, Vettukattil JJ, Stumper O, Wright JGC, Silove ED et al (2000) Balloon angioplasty in infants with aortic obstruction after the modified stage I Norwood procedure. *Am Heart J* 140:227–231
6. Cho GY, Song JK, Park WJ, Han SH, Choi SH, Doo YC et al (2005) Mechanical dyssynchrony assessed by tissue Doppler imaging is a powerful predictor of mortality in congestive heart failure with normal QRS duration. *J Am Coll Cardiol* 46:2237–2243
7. Choussat A, Fontan F, Besse P (1977) Selection criteria for the Fontan procedure. In: Anderson RH, Shinebourne EA (eds) *Paediatric cardiology*. Churchill Livingstone, Edinburgh, Scotland, pp 559–566
8. Colliou O, Thompson T, Jensen M, Zdeblick M, Thompson A, Costello B et al. (2006) Patent application title: Deployable epicardial electrode and sensor array
9. Deal BJ, Mavroudis C, Backer CL (2007) Arrhythmia management in the Fontan patient. *Pediatr Cardiol* 28:448–456
10. Driscoll DJ (2007) Long-term results of the Fontan operation. *Pediatr Cardiol* 28:438–442
11. Dubin AM, Janousek J, Rhee E, Strieper MJ, Cecchin F, Law IH et al (2005) Resynchronization therapy in pediatric and congenital heart disease patient: an international multicenter study. *J Am Coll Cardiol* 46:2277–2283
12. Hellenbrand WE, Allen HD, Golinko RJ, Hagler DJ, Lutin W, Kan J (1990) Balloon angioplasty for aortic recoarctation: results of valvuloplasty and angioplasty of congenital anomalies registry. *Am J Cardiol* 65:793–797
13. Holzer RJ, Chisolm JL, Hill SL, Cheatham JP (2008) Stenting complex aortic arch obstructions. *Catheter Cardiovasc Interv* 71:375–382
14. Ichikawa H, Yagihara T, Kishimoto H, Isobe F, Yamamoto F, Nishigaki K et al (1995) Extent of aortopulmonary collateral blood flow as a risk factor for Fontan operations. *Ann Thorac Surg* 59:433–437
15. Ishibashi N, Aoki M, Watanabe M, Kakajima H, Aotsuka H, Fujiwara T (2008) Risk factor of interim failure and early detection of the high-risk patients with functional single ventricle after Blalock–Taussig shunt. *J Card Surg* 23:488–492
16. Jacobs ML, Pourmoghadam K (2007) Thromboembolism and the role of anticoagulation in the Fontan patient. *Pediatr Cardiol* 28:457–464
17. Janousek J, Tomek V, Chaloupecký V, Reich O, Gebauer RA, Kautzner J et al (2004) Cardiac resynchronization therapy: a novel adjunct to the treatment and prevention of systemic right ventricular failure. *J Am Coll Cardiol* 44:1927–1931
18. Kanter KR, Vincent RN, Raviele AA (1999) Importance of acquired systemic-to-pulmonary collaterals in the Fontan operation. *Ann Thorac Surg* 68:969–975
19. Khairy P, Fournier A, Thibault B, Dubuc M, Therien J, Vobecky SJ (2006) Cardiac resynchronization therapy in congenital heart disease. *Int J Cardiol* 109:160–168
20. Khairy P, Fernandez SM, Mayer JE, Triedman JK, Wals EP, Lock JE et al (2008) Long-term survival, modes of death, and predictors of mortality in patients with Fontan surgery. *Circulation* 117:85–92
21. Lechner E, Gitter R, Mair R, Pinter M, Schreier-Lechner E, Vondrys D et al (2008) Aminoterminal brain peptide levels in children and adolescents after Fontan operation correlate with congestive heart failure. *Pediatr Cardiol* 29:901–905
22. Mahle WT, Wernovsky G, Bridges ND, Linton AB, Paridon SM (1993) Impact of early ventricular unloading on exercise performance in preadolescents with single-ventricle Fontan physiology. *J Am Coll Cardiol* 34:1637–1643
23. Moskowitz WB, Schieken RM, Mosteller M, Bossano R (1990) Altered systolic and diastolic function in children after "successful" repair of coarctation of the aorta. *Am Heart J* 120:103
24. Muhll IV, Liu P, Webb G (2004) Applying standard therapies to new targets: the use of ACE inhibitors and B-blockers for heart failure in adults with congenital heart disease. *Int J Cardiol* 97:25–33
25. Nagendran J, Archer SL, Soliman D, Gurtu V, Moudgil R, Harmony A et al (2007) Phosphodiesterase type 5 is highly expressed in the hypertrophied human right ventricle, and acute inhibition of phosphodiesterase type 5 improves contractility. *Circulation* 116:238–248
26. Odegard KC, Zurakowski D, DiNardo JA, Castro RA, McGowan FX Jr, Neufeld EJ et al (2009) Prospective longitudinal study of coagulation profiles in children with hypoplastic left heart syndrome from stage I through Fontan completion. *J Thorac Cardiovasc Surg* 137:934–941
27. Ohye RG, Gomez CA, Goldberg CS, Graves HL, Devaney EJ, Bove EL (2004) Tricuspid valve repair in hypoplastic left heart syndrome. *J Thorac Cardiovasc Surg* 127:465–472
28. Pass RH, Hsu DT, Garabedian CP, Schiller MS, Jayakumar KA, Hellenbrand WE (2002) Endovascular stent implantation in the pulmonary arteries of infants and children without the use of a long vascular sheath. *Catheter Cardiovasc Interv* 55:505–509
29. Piran S, Veldtman G, Siu S, Webb GD, Liu PP (2002) Heart failure and ventricular dysfunction in patients with single or systemic right ventricles. *Circulation* 105:1189–1194

30. Ralph-Edwards AC, Williams WG, Coles JC, Rebeyka IM, Trusler GA, Freedom RM (1995) Reoperation for recurrent aortic coarctation. *Ann Thorac Surg* 60:1303–1307
31. Sasaki T, Takahashi Y, Ando M, Wada N (2007) Edge-to-edge tricuspid valve repair in hypoplastic left heart syndrome. *Gen Thorac Cardiovasc Surg* 55:505–507
32. Senzaki H, Masutani S, Kobayashi J, Kobayashi T, Sasaki N, Asano H et al (2002) Ventricular afterload and ventricular work in Fontan circulation. Comparison with normal two-ventricle circulation and single-ventricle circulation with Blalock–Taussig shunts. *Circulation* 105:2885–2892
33. Senzaki H, Kyo S, Matsumoto K, Asano H, Masutani S, Ishido H et al (2004) Cardiac resynchronization therapy in a patient with single ventricle and intracardiac conduction delay. *J Thorac Cardiovasc Surg* 127:287–288
34. Sojak V, Mazic U, Cesen M, Schrader J, Danojevic N (2008) Cardiac resynchronization therapy for the failing Fontan patient. *Ann Thorac Surg*; 85:2136–2138
35. Spicer RL, Uzark KC, Moore JW, Mainwaring RD, Lamberti JJ (1996) Aortopulmonary collateral vessels and prolonged pleural effusions after modified Fontan procedures. *Am Heart J* 131: 1164–1168
36. Stern HJ (2010) Aggressive coiling of aortopulmonary collaterals in single-ventricle patients is warranted. *Pediatr Cardiol* 31: 449–453
37. Stern HJ, Baird CW (2009) A premounted stent that can be implanted in infants and re-dilated to 20 mm: introducing the Edwards Valeo lifestent. *Catheter Cardiovasc Interv* 74:905–912
38. Szabo G, Buhmann V, Graf A, Melnitschuk S, Bährle S, Vahl CF et al (2003) Ventricular energetics after the Fontan operation: contractility-afterload mismatch. *J Thorac Cardiovasc Surg* 125:1061–1609
39. Tomaske M, Gerritse B, Kretzers L, Pretre R, Dodge-Khatami A, Rahn M et al (2008) A 12-Year experience of bipolar steroid-eluting epicardial pacing leads in children. *Ann Thorac Surg*; 85:1704–1711
40. van den Bosch AE, Roos-Hesselink JW, van Domburg R, Bogers AJJC, Simoons ML, Meijboom FJ (2004) Long-term outcome and quality of life in adult patients after the Fontan operation. *Am J Cardiol* 93:1141–1145
41. Vitarelli A, Conde Y, Cimino E, D’Orazio S, Stellato S, Battaglia D et al (2008) Assessment of ascending aorta distensibility after successful coarctation repair by strain Doppler echocardiography. *J Am Soc Echocardiogr* 21(6):729–736
42. Vogel M, Schmidt MR, Kristiansen SB, Cheung M, White PA, Sorensen K et al (2002) Validation of myocardial acceleration during isovolumic contraction as a novel noninvasive index of right ventricular contractility: comparison with ventricular pressure-volume relations in an animal model. *Circulation* 105: 1693–1699
43. Yan RT, White M, Yan AT, Yusuf S, Rouleau JL, Maggioni AP et al (2005) Usefulness of temporal changes in neurohormones as markers of ventricular remodeling and prognosis in patients with left ventricular systolic dysfunction and heart failure receiving either candesartan or enalapril or both. *Am J Cardiol* 96(5): 698–704
44. Yetman AT, Nykanen D, McCrindle BW, Sunnegardh J, Adatia I, Freedom RM et al (1997) Balloon angioplasty of recurrent coarctation: a 12-year review. *J Am Coll Cardiol* 30:811–816
45. Zaidenweber CM, Kim DW, Vincent RN (2006) Right ventricular outflow tract and pulmonary artery stents in children under 18 months of age. *Catheter Cardiovasc Interv* 69:23–27