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Early mortality and concomitant procedures related to Fontan conversion: Quantitative analysis☆

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ABSTRACT

Background: The Fontan palliation is associated with numerous complications during long-term. The Fontan conversion operation has been advocated as an option to avoid some of these problems by converting classical Fontan types to modern forms of the circulation. Early mortality of Fontan conversion, however, remains unclear as available reports include limited numbers of patients and the results are heterogeneous.
Methods: We reviewed all original articles from 1994 to 2016 reporting Fontan conversion operations. Reports were analysed with specific reference to patient demographics, patient number, concomitant arrhythmia surgery, pacemaker implantation and early mortality.
Results: Overall, 37 Fontan conversion studies with a total of 1182 patients were analysed, including 35 single-centre studies and 2 registers. In the 35 single-centre studies the average age at the time of conversion was 21.6 years (range 10.2–30.9 years). Concomitant arrhythmia operation was performed in 71.6% of patients and concomitant pacemaker implantation procedure was performed in 59.3% of patients. Early mortality varied greatly between publications ranging from 0 to 21%. Based on a random and a fixed effect model mean mortality was 5.3% and 6.2%, respectively. Lower mortality was observed in series including younger patients at the time of conversion (average age < 20 years, 4.6%) and in the highest volume centre (1.4%).
Conclusion: Fontan conversion carries a substantial mortality risk. However, results vary between centres. Overall, the combination with arrhythmia surgery seems to be associated with lower early mortality especially when patients are referred at an earlier age and are treated at highly experienced centres.

1. Introduction

The Fontan procedure represents a milestone in the management of patients with functionally univentricular hearts [1,2]. It remains a prime example of major innovation with uncertain long-term consequences in the field of congenital heart disease to this day. The procedure enables satisfactory mid- to long-term survival of even the most complex heart defects with functionally univentricular circulation - such as tricuspid atresia or hypoplastic left heart syndrome - that previously resulted in death during early childhood in the vast majority of patients. Even though reasonable survival has been reported decades after the Fontan procedure, the procedure itself remains palliative in nature and is inherently associated with a high and persistent risk of arrhythmias, [3,4] thromboembolic events, [5–7] exercise intolerance [8] and premature death. In fact, Fontan patients as a group had the highest standardized mortality ratio and the highest 5-year risk of death in adulthood among all groups of adult congenital heart disease patients included in a recent study [9].

The Fontan type palliation cannot be considered a uniform type of operation as the procedure has gone through an evolution since its first description in 1971. Although, all types of Fontan procedure divert the systemic venous return without the interposition of a functional subpulmonary ventricle to the pulmonary circulation, they differ greatly in the way the right atrium is utilized. Whereas, the atrio-pulmonary connection (APC) type of Fontan operation incorporates the right atrium in the Fontan circulation, surgical modifications (i.e. the lateral tunnel [LT] or extracardiac conduit [EC]) partially or complete exclude this chamber from the Fontan pathway. The rationale for the development of these modified techniques was both theoretical (improved hydrodynamics and lower energy loss in the venous pathway) [10,11] and from previous experience of high complication rates with the APC

☆ "Authors take responsibility for all the aspects of the reliability and freedom from bias of the data presented and their discussed interpretation."

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Table 1
Summary of all included single centre studies [17,19,28,30–60] as well as of the registry studies based on the datasets from the Society of Thoracic Surgeons Congenital Heart Surgery Database (STS-CHSD) and the Congenital Heart Surgery Databases (CHSDBs) of the European Congenital Heart Surgeons Association (ECHSA) [21,26].
/indicates data not provided in the respective study. *Nb = number. OP = operation. PM = pacemaker.*

Number	Author	Centre	Reference	Year	Nb of patients	Early mortality Nb	Early mortality %	Era	Average age of conversion	Arrhythmia Op %	PM implantation %
1	Mavroudis C	Chicago IL, USA	J Thorac Cardiovasc Surg 1998	1998	14	1	7,0%	1992–1997	14	79,0%	71,0%
2	Mavroudis C	Chicago IL, USA	J Thorac Cardiovasc Surg 2001	2001	40	0	0,0%	1994–2001	18,7	100,0%	95,0%
3	Deal BJ	Chicago IL, USA	Ann Thorac Surg 2016	2016	140	2	1,4%	1994–2012	23,2	99,0%	98,6%
4	Said SM	Rochester, Minnesota, USA	Ann Thorac Surg 2014	2014	70	10	14,0%	1994–2011	23,2	77,0%	57,0%
5	Pundi KN	Rochester, Minnesota, USA	J Am Coll Cardiol 2015	2015	117	10	9,0%	1973–2012	/	/	/
6	Kreutzer J	Boston, MA, USA	J Thorac Cardiovasc Surg 1996	1996	8	1	12,5%	1990–1994	16,5	0,0%	12,5%
7	Takahashi K	Boston, MA, USA	International J Card 2009	2009	40	5	12,5%	1990–2006	19	53,0%	45,0%
8	Petko M	Philadelphia, USA	Euro J of Cardio-thoracic Surgery 2003	2003	13	1	7,7%	1995–2001	/	38,5%	0,0%
9	Morales DLS	Houston, Texas, USA	Ann Thorac Surg 2005	2005	35	0	0,0%	1997–2004	19,2	80,0%	83,0%
10	Weinstein S	Columbus, Ohio, USA	Semin Thorac Cardiovasc Surg 2005	2005	15	2	13,0%	1999–2004	25	100,0%	93,0%
11	Kao JM	Los Angeles, CA, USA	Ann Thorac Surg 1994	1994	3	0	0,0%	1993	14,9	0,0%	0,0%
12	McElhinney DB	San Francisco, CA, USA	Ann Thorac Surg. 1996	1996	7	1	14,3%	1992–1995	20	0,0%	0,0%
13	Aboulhosn J	Los Angeles, CA, USA	Congenit Heart Dis. 2010	2010	27	2	7,4%	1993–2009	30	78,0%	41,0%
14	Abella RF	Modena, IT	G Ital Cardiol 1998	1998	9	0	0,0%	1994–1997	21,5	0,0%	0,0%
15	Conte S	Leuven, Belgium	Cardiovasc Surg 1999	1999	7	1	14,3%	1994–1998	15,6	14,0%	14,0%
16	van Son JA	Leipzig, Germany	Eur J Cardiothor Surg 1999	1999	18	2	11,0%	1994–1999	15,1	0,0%	0,0%
17	Marcelletti CF	Modena, IT	J Thorac Cardiovasc Surg 2000	2000	31	2	6,5%	1992–1999	19,9	26,0%	19,0%
18	Agnoletti G	Bergamo, IT	Heart 2003	2003	11	1	9,1%	1999–2000	20,9	100,0%	100,0%
19	Sheikh AM	Southampton, United Kingdom	J Thorac Cardiovasc Surg 2004	2004	15	0	0,0%	1997–2002	19,7	73,0%	73,0%
20	Erek E	Istanbul, Turkey	J Thorac Cardiovasc Surg 2006	2006	5	0	0,0%	1997–2005	10,2	100,0%	0,0%
21	Giardini A	Bologna, IT	International Journal of Cardiology 2006	2006	6	0	0,0%	/	19	33,0%	100,0%
22	Sridhar A	Milan, IT	Cardiology in the Young 2011	2011	15	2	13,3%	2002–2009	26,2	100,0%	47,0%
23	Ono M	Munich, Germany	Thorac Cardiovasc Surg 2015	2015	15	1	6,7%	2006–2014	30,9	100,0%	100,0%
24	Kim WH	Seoul, South Korea	European Journal of Cardio-Thoracic Surgery 2005	2005	16	0	0,0%	1996–2004	17	81,0%	44,0%
25	Kwak JG	Seoul, South Korea	Cardiac Surg 2009	2009	18	1	5,6%	1996–2007	/	89,0%	61,0%
26	Jang WS	Seoul, South Korea	Euro J of Cardiothor Surg 2014	2014	31	0	0,0%	1996–2011	19	83,9%	74,2%
27	Kawahira Y	Osaka, Japan	Ann thorac Surg 2001	2001	4	0	0,0%	/	21	100,0%	0,0%
28	Koh M	Osaka, Japan	J Thorac Cardiovasc Surg 2007	2007	10	0	0,0%	/	22	100,0%	0,0%
29	Fujita S	Tokyo, Japan	Japanese College of Cardiology 2009	2009	14	3	21,0%	/	/	43,0%	/
30	Hoashi T	Osaka, Japan	Kyobu Geka 2010	2010	7	0	0,0%	1999–2008	20,3	100,0%	0,0%
31	Hiramatsu T	Tokyo, Japan	Eur J Cardiothorac Surg 2011	2011	38	3	7,9%	1992–2010	25,8	58,0%	58,0%
32	Terada T	Nagoya-city, Aichi, Japan	Asian Cariovasc&Thoracic Annals 2014	2013	25	0	0,0%	2004–2012	21	96,0%	24,0%
33	Setty SP	Auckland, New Zealand	Ann Thorac Surg 2002	2002	6	0	0,0%	1997–2001	22,8	100,0%	100,0%
34	Poh CL	Australia & New Zealand	European Journal of Cardio-Thoracic Surgery 2016	2016	39	4	10,3%	1990–2014	23,8	95,0%	41,0%
35	Park HK	Seoul, South Korea	Interactive CardioVascular and Thoracic Surgery 2016	2016	21	0	0,0%	1998–2013	17,9	38,1%	14,3%
36	Fuller MS	STS Congenital Hear Surgery Database	Ann Thorac Surg 2015	2015	155	/	10,30%	2000–2013	26.1?	/	/
37	van Melle JP	ECHSA European Congenital Heart Surgeons Association	Heart 2016	2016	137	15	10,90%	1986–2012	/	51,60%	/

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