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Is vaginal delivery or caesarean section the safer mode of delivery in patients with adult congenital heart disease?

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Abstract

A best evidence topic in cardiac surgery was written according to a structured protocol. The question addressed was: is vaginal delivery or caesarean section (CS) the safer mode of delivery in patients with adult congenital heart disease? Of the 119 studies, 13 papers represented the best evidence on the topic. Recommendations are based on 29 262 patients. Those having undergone successful corrective or palliative cardiac surgery for congenital heart disease, in addition to patients with unoperated congenital heart disease are a high-risk obstetric population. Heart disease is a leading cause of maternal mortality in the USA and the UK. Traditionally, CS was regarded as the mode of delivery of choice for high-risk patients, but growing experience in this field has now made this advice appear controversial. Patients are stratified into high- and low-risk, depending on the degree of heart failure symptoms [New York Heart Association (NYHA) class]. All studies demonstrated adverse outcomes in ACHD patients compared with normal age-matched controls. This pertained to a higher overall risk of maternal cardiac death, neonatal death, preterm birth, fetal growth restriction and longer hospital stay. On univariate regression analysis, the variables that imparted the highest risk to mother and foetus, were right ventricular failure, pulmonary regurgitation and pulmonary hypertension ($P < 0.001$). Induction of labour was deemed safe and was not associated with higher CS rates. There was no increase in maternal or neonatal complications in patients who were NYHA class I and II at labour. Patients who were NYHA class III and IV at labour had higher complication rates with adverse feto-maternal outcomes ($P < 0.0001$) and longer intensive care unit and hospital stay (Spearman's correlation 0.326, $P = 0.007$). The largest cohort from the USA (26 973 ACHD births) demonstrated that ventricular septal defect was associated with the highest risk of maternal death and complications ($P < 0.05$). The data would indicate that patients NYHA class I and II symptoms are suitable for VD. For most NYHA III and IV patients a trail of labour is safe with expedited delivery under good analgesic control as dictated by obstetric needs. Due to high complication risks, CS may be indicated in a proportion of patients.

Keywords: Adult congenital heart disease • Normal delivery • Vaginal delivery • Caesarean section • Maternal mortality

INTRODUCTION

A best evidence topic was constructed according to a structured protocol. This is fully described in ICVTS [1].

THREE-PART QUESTION

In which patients with [adult congenital heart disease] is [vaginal delivery] safer compared to a [caesarean section (CS)]?

CLINICAL SCENARIO

A 35-year old nulliparous woman wishes to start a family. She had an operation when she was a baby to correct a ventricular septal defect (VSD). She has been well since her operation. She is not on any medication. She has normal exercise tolerance. She is keen to have a pregnancy and normal delivery. She asks for your opinion, as to whether her heart is likely to tolerate the stress of labour and normal delivery.

SEARCH STRATEGY

Medline search 1950–December 2012 was performed using OVID interface [heart defects, congenital/ and adult] OR [adult congenital heart disease.mp] AND [delivery, obstetric] OR [labour, obstetric] OR [extraction, obstetric] OR [vacuum extraction, obstetrical] OR [obstetric labour complications] OR [CS].

SEARCH OUTCOME

One hundred and nineteen studies were identified, of these, 13 papers represented the best evidence on the topic (Table 1). Case reports and studies with <25 patients were excluded. Studies with overlapping patient groups from the same institutions or those with unclear clinical outcomes were also excluded.

Table 1: Summary of best evidence

Author, date and country Study type (level of evidence)	Patient group	Outcome	Key results	Comments
Roos-Hesselink *et al.* (2012), Eur Heart J, Netherlands [2]	60 hospitals, 28 countries (2007–2011) *n* = 1321 ACHD 872 (66%)	CS rate	NYHA I 76%, II 21%, III 1.3% and IV 1.5%	Significantly higher CS rate was attributed to prematurity
Multicentre prospective International registry (level 1c)	Non-ACHD CS rate 23% [2]	Maternal mortality	CS rate 38% ($P < 0.001$)	Mortality: ACHD 0.5% Cardiomyopathy 2.4% Valvular heart disease 2.1% Ischaemic heart disease 0%
		Neonatal death	Maternal mortality 0.5 vs 0.007% ($P < 0.001$)	
		Heart failure	Neonatal death 0.6 vs 0.4% ($P < 0.001$)	
		Prematurity	Heart failure 8 vs 0% ($P < 0.001$) Preterm birth 13 vs 8% ($P < 0.001$)	
Siu *et al.* (2001), Circulation, Canada [3]	1994–1999 445/562 (74%) consecutive ACHD patients	CS rate	NYHA I and II 96%; NYHA III 4%	Symptomatic high-risk patients should have cardiac intervention before pregnancy. Poor NYHA class, cyanosis, myocardial dysfunction, arrhythmia and heart failure/stroke patients need management in tertiary centre
Prospective multicentre national registry (level 1c)	National CS rate (2008) 26.5% [16]	Cardiac complications	CS rate 27%	
		Neonatal outcomes	Live birth rate 98%; 96% CS performed for obstetric indications; maternal cardiac status was the indication in 4%	
			Independent risk factors of adverse maternal outcome:	
			NYHA >II or cyanosis (OR 6, $P < 0.03$); arrhythmia (OR 6, $P < 0.001$); poor LVEF (OR 11, $P < 0.001$); left heart obstruction (OR 6 $P < 0.001$)	
			Independent risk factors of adverse neonatal outcome:	
			NYHA >II or cyanosis (OR 3, $P < 0.03$); left heart obstruction (OR 2 $P < 0.04$); heparin/warfarin (OR 3 $P < 0.0093$)	
Karamlou *et al.* (2011), Ann Thorac Surg, USA [4]	Epidemiological review (1998–2007) *n* = 39.9 million births	CS rate	Complications higher for ACHD compared with age-matched women:	Authors conclude that there is no triage of patients with even relatively 'simple' ACHD lesions
Retrospective multicentre national review (level 2a)	ACHD births = 26 973 (0.07%)	Obstetric outcomes	ACHD CS rate 33.6% ($P < 0.001$)	Improved education and triage are needed to improve outcomes
	Non-ACHD CS rate 27% [4]	Maternal mortality	Surgically assisted vaginal delivery (11.8 vs 7.9%)	
		Cardiac complications	Induction (37 vs 33%)	
		Prematurity	Maternal mortality 18-fold higher in ACHD (*n* = 25; 0.09% vs *n* = 2119; 0.005%; $P < 0.001$)	
			Cardiac complications (2.3 vs 0.2%, $P < 0.001$)	
			Preterm delivery (10 vs 7%, $P < 0.001$)	
			Stillbirth (0.8 vs 0.7%)	

Continued

Table 1: (*Continued*)

Author, date and country Study type (level of evidence)	Patient group	Outcome	Key results	Comments
			ACHD patients delivered at teaching hospitals (58 vs 45%; $P < 0.001$)	
Hidano *et al.* (2011), Int J Obstet Anesth, Japan [5] Retrospective cohort study (level 2b)	7 years retrospective study $n = 128$ women with 151 deliveries National CS rate (2008) 17.4% [16]	CS rate Maternal and neonatal mortality Neonatal morbidity Cardiac complications	All NYHA I and II. ACHD CS rate: 67/151 (44%) - a third for maternal cardiac complication Vaginal birth: 84/151 (56%) Assisted vaginal birth: 37/84 (44%) General anaesthesia: 17/68 (25%) No maternal deaths; 2 neonatal deaths (one vaginal; one caesarean delivery) Neonatal complications after CS 25/68 (37%); transient tachypnoea 5; SGA 8; prematurity 20 Maternal cardiac events after vaginal birth 1/84 (1%); after CS 10/67 (15%) Neonatal complications: vaginal delivery 11/84 (13%); CS 25/67 (37%) 23 Elective CS: higher rate of maternal cardiac (35%) and neonatal (65%) complications	There was a low overall incidence of maternal and neonatal mortality. Pregnancy with ACHD was associated with significant maternal cardiac and neonatal complications 15/23 (65%) neonatal complications where highest when CS was done for maternal cardiac indications. Compared to: obstetric 6/28 (21%); foetal 4/16 (25%) High risk lesions: Corrected or uncorrected transposition of great vessels, Fallots tetralogy, VSD; PDA untreated Highest risk lesions: Eissenmengers developed in uncorrected Tetralogy of Fallot; Marfan's with aortic dilation >40 mm
Goldszmidt *et al.* (2010), Int J Obstet Anesth, Canada [6] Retrospective cohort study (level 2b)	1986–2004 $n = 276/522$ ACHD patients National CS rate (2008) 26.3% [16]	Operative birth Prematurity	268/276 (97%) NYHA class I and II; 7 (3%) NYHA class III and IV General anaesthesia: caesarean delivery (AOR 0.74; 95% CI 9.5); complex congenital heart lesion (OR 2.3; 95% CI 1.0); prematurity (OR 1.3; 95% CI 1.1) When adjusted for multiple births complex congenital cardiac defects were not associated with general anaesthetic (AOR 2.8; 95% CI 0.76, 10.1)	Pregnant women with ACHD require an organized program for labour and delivery
Ouyang *et al.* (2010), Int J Cardiol, USA [7] Retrospective cohort study (level 2b)	1998–2005 $n = 112$ pregnancies; $n = 92$ to >20 weeks; $n = 65$ ACHD National CS rate (2008) 30.3% [16]	CS rate Neonatal outcomes No Valsalva vs Valsalva	All NYHA I and II CS rate 31.5% 62 pregnancies reached 2nd stage labour 90 live births NICU admission ($n = 21$); SGA ($n = 18$); prematurity ($n = 19$, of which 4/19 were <28 weeks), 1 death No Valsalva ($n = 45$): all vaginal births; 82.2% instrumental; 8/45 PPH; 7/45 3rd/4th degree tear; 1	All CS for obstetric indications, except of a combined case of aortic root surgery and CS; for coarctation with 6 cm aortic root dilatation Beta-blockers associated with SGA ($P = 0.001$) 8/90 (8.9%) neonates diagnosed with congenital heart disease

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Table 1: (*Continued*)

Author, date and country Study type (level of evidence)	Patient group	Outcome	Key results	Comments
			cardiac event (D-transposition of the great arteries, had CCF 2 days post-partum) Valsalva ($n = 17$): 16 vaginal births, 11.7% instrumental; 1 CS arrest of descent; no PPH, no 3rd/4th degree tears Duration of 2nd stage was longer (60 vs 21 min) ($P = 0.075$)	
Curtis *et al.*, (2009), Int J Cardiol, UK [8] Retrospective cohort study (level 2b)	1999–2005 $n = 101$ patients with 131 pregnancies National CS rate (2008) 22% [16]	CS rate Cardiac complications	NYHA III and IV (7%): Vaginal delivery 21.4% (3/14); CS 78.6% (11/14) 6 cardiac indication; 5 obstetric indication Cardiac intervention rate in pregnancy 13/101 (12.9%)	There is a sustained increase in ACHD pregnancies Preconception advice and the follow-up needs to be at a tertiary hospital High risk lesions for cardiac events include: severe AR/MR (deterioration in NYHA); poor LV function; congenital complete heart block, dilated cardiomyopathy
Wasim *et al.* (2008), J Pak Med Assoc, Pakistan [9] Retrospective 5-year cohort study (level 2b)	$n = 17\,056$ births 160 cardiac patients ACHD 28/160 (17.5%) National CS rate (2008) 7.3% [16]	CS rate Neonatal outcomes Maternal mortality	57% NYHA class I and II 43% NYHA class III and IV CS rate 29% Neonatal mortality 10/160 (6.2%) Maternal mortality (3.8%)	NYHA III and IV key determinant of adverse feto-maternal outcome ($P < 0.0001$)
Meng *et al.* (2007), Zhonghua Fu Chan Ke Za Zhi, China [10] Retrospective cohort study (level 2b)	1995–2007 45 ACHD patients with pulmonary hypertension National CS rate (2008) 25.9% [16]	CS rates Maternal mortality	NYHA I and II 97%; NYHA III and IV 3% Overall CS rate 78% (35/45) Maternal mortality 4% (2/45) Vaginal delivery 22% (10/45) CCF 24.4% (11/45)	Mild pulmonary HTN: CS rate 76% (22/29) Term delivery 93% (27/29) Prematurity 3% (1/29) Abortion 3% (1/29) Moderate pulmonary HTN: CS rate 75% (6/8) Term delivery (62.5%) 5/8 Prematurity (37.5%) 3/8 Severe pulmonary hypertension: CS rate 7/8 (87.5%) Term delivery 5/8 (62.5%) Prematurity 2/8 (25%) Latrogenic abortion 1/8 (12.5%)
Sidlik *et al.* (2007), J Matern Fetal Neonatal Med, Israel [11] Retrospective cohort study (level 2b)	1989–2002 67 ACHD patients 156 deliveries National CS rate 19.1% [16]	CS rate Cardiac complications Neonatal outcomes	NYHA I and II 99.1% CS rate 13% Lesions include: VSD (43.2%), bicuspid valve (20.8%) and aortic regurgitation (17.9%) ACHD independent risk factor of neonatal malformations ($n = 34$) (OR 2.1, 95%, CI 1.18–3.72)	No difference in maternal and neonatal outcomes between modes of deliveries CS rates similar to normal population. Congenital heart disease in baby 12/67 (17.9%) No reported mortalities.
Khairy *et al.* (2006), Circulation, USA [12]	1998–2004 $n = 53$ ACHD patients with 90 pregnancies	CS rate	All NYHA I and II CS rate 17/72 (23.6%); 20 SVD;	CS rate similar to background population ACHD was associated with significant

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Table 1: (*Continued*)

Author, date and country Study type (level of evidence)	Patient group	Outcome	Key results	Comments
Retrospective cohort study (level 2b)	National CS rate (2008) 30.3% [16]	Predictors of adverse perinatal events	22 forceps; 13 ventouse Independent predictors of primary cardiac events in pregnancy: Baseline NYHA ≥2 (OR 5.4, $P = 0.032$); history of heart failure (OR 15.5, $P = 0.02$); smoking (OR 15.6, $P = 0.002$); severe pulmonary regurgitation, or depressed subpulmonary ventricular EF (OR 9, $P = 0.01$) Maternal predictors of neonatal events: Subaortic ventricular outflow tract gradient >30 mmHg (OR 7.5, $P = 0.01$); smoking (OR 8, $P = 0.01$); symptomatic arrhythmia in pregnancy (OR 5.2, $P = 0.03$)	foetal and maternal cardiac complications No reported maternal mortalities Maternal cardiac events complicated 19.4% pregnancies (16.7% pulmonary oedema, 2.8% arrhythmias)
Boyle *et al.* (2003), Int J Obstet Anesth, Australia [13]	1993–1997 $n = 78$ deliveries in 68 women	CS rate Maternal mortality	NYHA I 41%, NYHA II 28% NYHA III 22%; NYHA IV 9%	2 mortalities (1. Known Eisenmenger's, with pulmonary hypertension. She had induced labour for cardiac indications, and vaginal delivery at 34 weeks. Died 3 days post-partum despite full cardiac management. 2. Severe mitral heart disease with pulmonary hypertension, had vaginal delivery at 38 weeks. Died 6 months later after open mitral valve replacement.)
Retrospective cohort study (level 2b)	ACHD 48/68 (70.5%) Rheumatic 17/68 (25%) Ischaemic 2/68 (2.9%) Illicit drug use 1/68 (1.5%) National CS rate (2008) 31% [16]	Cardiac complications	CS rate 28% (22/78) Maternal mortality 2.9% (2/68) NYHA III and IV had higher complication rate, longer ITU/ hospital stay ($P = 0.007$)	
McFaul *et al.* (1988), Br J Obstet Gynaecol, UK [14]	1970–1983 161 ACHD patients	Maternal mortality Cardiac complications	All maternal deaths occurred in NYHA III/IV. Heart failure in 18% pregnancies antenatally	NYHA I/II patients can safely deliver vaginally. NYHA III/IV are at high risk of adverse outcomes
Retrospective cohort study (level 2b)		Perinatal mortality	Perinatal mortality was rare 19/1000	

AOR: adjusted odds ratio; CS: caesarean section; SVD: spontaneous vaginal delivery; SGA: small for gestational age; RDS: respiratory distress syndrome; IVH: intraventricular haemorrhage; OR: odds ratio; LVEF: left ventricular ejection fraction; NYHA: New York Heart Association; VSD: ventricular septal defect; ITU: intensive care unit.

Primary outcomes of interest were safe mode of delivery and mortality rates. Secondary outcomes of interest were instrumental delivery rates, CS rates, adverse maternal cardiac event and neonatal death.

RESULTS

Roos-Hesselink *et al.* [2] showed significantly higher maternal and neonatal mortality in ACHD compared with the normal population. The CS rate (38%) was significantly higher in ACHD ($P < 0.001$). ACHD had better outcomes compared with cardiomyopathy and valvular and ischaemic heart disease.

A Canadian prospective multicentre registry consisting of 74% ACHD patients [New York Heart Association (NYHA) I/II 96%] [3] had a CS rate in ACHD patients similar to the background population. Independent risk factors identified for adverse maternal outcomes were NYHA > II or cyanosis (OR 6, $P = 0.009$); arrhythmia (OR 6, $P < 0.001$, poor left ventricular ejection fraction (OR 11, $P < 0.001$); left heart obstruction (OR 6 $P < 0.001$).

Karamlou *et al.* [4] reported 26 973 ACHD births. ACHD had a significantly higher rate of CS and overall maternal and neonatal morbidity and mortality, compared with age-matched controls. Diagnoses include: VSD ($n = 4152$, 15%); aortic valve pathology ($n = 3412$, 12.7%); ostium secundum atrial septal defect (ASD) ($n = 3402$, 12.6%). VSD was associated with the highest risk of maternal death and complications ($P < 0.05$).

Hidano *et al.* [5] reported 7-year outcomes in 151 births in ACHD patients. The series consisted of NYHA I/II, and severe lesions were delivered electively by CS. The CS rate in ACHD was significantly higher than the national average, of which 25%

were done under general anaesthetic. Most complications occurred in the CS group (13% after VD and 37% after CS). Most of these adverse outcomes occurred in the elective caesarean group (35% maternal cardiac and 65% neonatal complications), done for maternal cardiac indications; this may also relate to a coexisting higher rate of foetal prematurity.

Goldszmith *et al.* [6] found a comparable CS and complication rates for all NYHA (97% NYHA I and II) severities undergoing labour. General anaesthetic was associated with prematurity and multiple births in the presence of complex congenital heart disease. Caesarean section, epidural and general anaesthesia rates are similar to those in the general obstetric population.

Ouyang *et al.* [7] examined the effect of avoiding the Valsalva manoeuvre, as this is a commonly given suggestion for ACHD. Valsalva was cardiovascularly safe. The routine practice of avoiding valsalva is associated with significantly longer second stage ($P = 0.075$) with higher rates of PPH ($P = 0.017$) and third/fourth degree tears ($P = 0.027$).

Curtis *et al.* [8] reviewed 101 patients, 93% NYHA I. In 3%, the defects became apparent during pregnancy. Outcomes in NYHA I/II were better compared with NYHA III/IV, ($P < 0.0001$). VD was the commonest mode of delivery even in NYHA class III/IV and was safe, with an overall CS rate 29% [8].

Wasim *et al.* [9] reported on 160 patients with heterogeneous cardiac lesions, including 28 ACHD patients. They had a large high-risk group (57% NYHA I/II and 43% NYHA III/IV), with a mortality of 3.8%. NYHA III and IV were the key determinants of adverse feto-maternal outcome ($P < 0.0001$).

Meng *et al.* found that increasing severity of pulmonary hypertension was associated with increasing preterm labour (7 vs 37.5%) [10]. There was no maternal mortality in NYHA I/II and mild to moderate pulmonary hypertension. All mortalities (4%) occurred in NYHA III/IV with severe pulmonary hypertension [10]. On univariate risk scoring history of CCF (odds ratio (OR) 15.5), NYHA ≥2 (OR 5.4), decreased right ventricle (RV) ejection fraction (OR 7.7) predicts poor outcomes. Independent predictors were decreased RV ejection fraction and/or severe pulmonary regurgitation (OR 9.0). In the presence of these variables, elective CS might be indicated.

Sadlik *et al.* [11] had a 13% CS rate that is comparable with the national average of 19%. They found ACHD to be an independent risk factor for neonatal malformations of 34/67 (50.7%). There were no differences in outcomes between mode of delivery for ACHD patients.

Khairy *et al.* [12] reported 90 pregnancies with univariate predictors of outcome being NYHA class >2 (OR 5.4) and decreased LV function (OR 7.7). Sixty-three percent of VD required instrumentation. 23.6% were CS and 76.4% VD, mode of delivery did not affect outcomes. Patients with impaired right ventricular function and severe pulmonary regurgitation had significantly poor outcomes.

Boyle *et al.* [13] reviewed 48 ACHD patients (70% NYHA III/IV) all of whom were offered vaginal birth. There were two maternal mortalities (2.5%). NYHA III/IV had a significantly larger number of ICU/CCU admissions (29.3%), vs 3.6% in the mild group ($P < 0.05$). Cardiac compromise occurred most during labour ($n = 73$), followed by the antenatal period ($n = 2$), the first week post-partum (1 patient), and 6 months after birth ($n = 2$). They had a CS rate of 28 vs 31% for the background population, when no elective caesareans were done for maternal cardiac indications.

McFaul *et al.* [14] reported outcomes over 13 years in 161 patients, recapitulating the finding that patients with NYHA III/IV symptoms accounted for all maternal deaths and a high incidence of perinatal complications. NYHA I/II could be safely delivered using VD.

CS is associated with higher blood loss, blood transfusion and greater overall fluctuations in haemodynamic status [15]. The patient has to be able to lie flat for at least 1 h during the procedure, which may not be possible in those with orthopnoea (NYHA III and IV). In centres where CS was only done for obstetric indications, i.e. with a CS rate similar to the background population, there was no significant difference in adverse outcomes between vaginal birth and CS [6, 11, 12]. Maternal outcomes are significantly affected by the nature of the cardiac lesion, and not by the mode of delivery [3, 4, 12]. Where possible, high-risk patients (such as NYHA III/IV, cyanosis, arrhythmia, heart failure, myocardial dysfunction, decreased RV ejection fraction, severe pulmonary valve regurgitation, subaortic ventricular outflow tract gradient >30 mmHg, VSD) should be identified and optimized prenatally [3, 4, 12]. Cardiac complications can occur at any time during pregnancy, birth or even up to 6 months after birth [13].

CLINICAL BOTTOM LINE

Vaginal birth is safe in patients with ACHD of all severities, and a higher CS rate does not translate into improved outcomes. The evidence suggests that a higher CS rate is in fact associated with an increased overall risk of adverse outcomes (including mortality) for the mother. Perinatal complications and maternal mortality are associated with NYHA III/IV symptoms. ACHD patients should be managed in a tertiary centre due to the potential high risk of adverse maternal and neonatal outcomes.

Conflict of interest: none declared.

BEST EVIDENCE TOPIC

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