Journal of Paediatrics and Child Health

doi:10.1111/jpc.12410

ORIGINAL ARTICLE

Improving diagnostic accuracy in the transport of infants with suspected duct-dependent congenital heart disease

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Aim: To identify factors that distinguish duct-dependent congenital heart disease (DDCHD) from non-DDCHD in newborn infants.

Method: A retrospective, cohort study. The Newborn Emergency Transport Service, Victoria (NETS) is a retrieval service for all inter-hospital neonatal transfers, and the Royal Children's Hospital, Melbourne (RCH) is a paediatric cardiac referral centre for the state of Victoria, Australia. All infants ≤10 days and ≥34 weeks gestation with suspected CHD and/or persistent pulmonary hypertension of the newborn (PPHN), transferred by NETS from non-tertiary neonatal units to RCH, over a 4-year period.

Results: Of 142 eligible infants, 81 had DDCHD and 61 had non-DDCHD, of whom 51 had PPHN. Diagnostic accuracy of DDCHD by the NETS team was 77%. Presence of a heart murmur, abnormal pulses, upper and lower limb blood pressure (BP) difference >10 mmHg, cardiomegaly, initial SpO_2 of <92%, PaO_2 <50 mmHg, and pre-post ductal SpO_2 difference >10% were significantly associated with DDHCD on univariate analysis. No single clinical finding was significantly associated with DDCHD on multivariate analysis. Labile SpO_2, abnormal lung parenchyma, mean BP <40 mmHg, pH <7.25, lactate >5 and FiO_2 >0.5 were significantly associated with non-DDCHD, but at multivariate analysis only labile SpO_2 and mean BP <40 mmHg were associated with non-DDCHD.

Conclusions: Clinical diagnosis of DDCHD outside of a cardiac centre is challenging. No single factor predicts DDCHD. Combined interpretation of clinical, physiological and x-ray findings may assist.

Key words: congenital heart disease; newborn infant; prostaglandin E_1; transport.

What is already known on this topic

1 Dilemmas exist for neonatal transport team to diagnose and manage congenital heart disease using limited information of clinical variables.
2 Distinguishing duct-dependent congenital heart disease from non-duct-dependent congenital heart disease has important therapeutic and transport implications.
3 Early use of prostaglandin E1 in hypoxic infants is beneficial and is not associated with identifiable adverse effect.

What this paper adds

1 A large number of infants with suspected DDCHD are delivered without antenatal diagnosis in non-tertiary settings.
2 Stratification of both clinical and investigative finding may assist in the diagnosis of duct-dependent congenital heart disease.
3 In the absence of a reliable single clinical discriminator, prostaglandin E1 should be used when duct-dependent congenital heart disease is suspected.

Despite advances in antenatal screening, a significant number of infants with congenital heart disease remain undiagnosed.[1] Prompt diagnosis and appropriate management are necessary to prevent deterioration and adverse outcome.[2] However, many infants are born in non-tertiary centres and require retrieval and transfer before a definitive echocardiographic diagnosis. The challenge for the referring and retrieving teams is to distinguish suspected duct-dependent congenital heart disease (DDCHD) from other causes of hypoxaemia, particularly persistent pulmonary hypertension of the newborn (PPHN).

Distinguishing DDCHD from non-DDCHD has important therapeutic and transport implications. Infants with DDCHD may require prostaglandin E_1 (PGE_1) to maintain ductal patency and prevent clinical deterioration.[3,4] In non-DDCHD, PGE_1 is not considered necessary and may produce unwanted side effects. Lewis *et al.* observed side effects in 20% of infants treated with PGE_1, of whom 12% developed respiratory depression.[5] Apnoea secondary to PGE_1 may be managed by intubation and ventilation, but at a cost of prolonging and complicating retrieval and transfer. Furthermore, infants with non-DDCHD (including

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Contributorship: All authors were responsible for planning and reporting. NG and NP conducted the data collection and analysis.
Funding: No funding provided/obtained for this study.
Conflict of interest: Nil.
Data sharing: No additional data.

Accepted for publication 11 June 2013.

predominantly PPHN) may benefit from pulmonary vasodilator therapy during transport whereas DDCHD may not.[6] There are also significant logistic and resource implications of distinguishing DDCHD and non-DDCHD; DDCHD usually requires transfer to a regional paediatric cardiac centre for ongoing management, whereas non-DDCHD may be appropriately managed in a neonatal intensive care unit without direct access to paediatric cardiology services.

This study aimed to identify factors which may improve differentiation of DDCHD and non-DDCHD in this group of challenging and vulnerable infants.

Methods

We conducted a retrospective cohort study of infants ≤10 days old and ≥34 weeks gestation with suspected DDCHD and/or PPHN transferred by the Newborn Emergency Transport Service, Victoria (NETS), from levels 1 and 2 special care nurseries to the regional paediatric cardiac centre at the Royal Children's Hospital, Melbourne (RCH), between 1 May 2007 and 31 May 2011.

Case identification

Cases were identified from combined database analysis (Fig. 1). Infants who received PGE_1 during transport were identified from the NETS database. Infants with suspected DDCHD or PPHN referred to the Cardiology service at RCH were identified from the cardiac database. Final diagnosis was obtained from the cardiac database based on echocardiographic findings. Infants were subdivided into DDCHD and non-DDCHD groups. The latter were sub-classified as PPHN or non-PPHN. Infants were also classified according to whether they received PGE_1.

Exclusions

Infants with life-threatening extra-cardiac anomaly, antenatal or postnatally diagnosed CHD, and those transferred from other tertiary centres were excluded.

Retrieval and stabilisation

In Victoria, all newborn infants transferred from levels 1 and 2 special care nurseries are retrieved and stabilised by NETS. The NETS team consists of a neonatal nurse and neonatal specialist trainee, supported by a NETS consultant. After initial assessment by NETS team, diagnosis and management plan are reviewed in a recorded teleconference with attending neonatologist and if necessary paediatric cardiologist may participate. Transfers are categorised based on clinical criteria, as 'time critical', 'primary urgent' and 'non-urgent', with an intention to depart the NETS base within 15, 25 or 60 min, respectively.

Data collection

Demographic and transport data were recorded. The clinical and laboratory findings recorded on initial NETS team assessment were: cyanosis, heart murmur, abnormal pulses (including reduced volume), labile saturations (max-min difference >10%), presence of pre-post ductal saturation difference >10%, upper and lower limb blood pressure (BP) difference >10 mmHg, mean BP, SpO2 (post-ductal), blood gas data, chest X-ray (CXR) findings (abnormal parenchyma and/or cardiomegaly). Use of PGE_1, respiratory and cardiovascular therapies were recorded.

Statistical analysis

Data were analysed using SPSS v18 (SPSS Inc., Chicago, IL, USA). Summary statistics were used to describe the population and subgroups. Comparison between groups was performed using χ^2 for categorical variables and *t*-test or non-parametric tests for continuous variables.

Ethics approval

The study was approved by the Research and Ethics committee at the RCH, Melbourne.

Results

One hundred forty-two eligible infants were identified (Fig. 1). Eighty-one infants had a final diagnosis of DDCHD (DDCHD Group). Of the 61 infants with non-DDCHD, 51 had PPHN (PPHN Group). Of the remaining 10 non-DDCHD infants, final diagnoses were severe sepsis (n = 5), minor congenital heart disease (CHD) (n = 2), cardiomyopathy (n = 1), pneumothorax (n = 1) and normal infant (n = 1). Diagnostic accuracy of DDCHD by transport team was 77%, with a sensitivity of 90% and positive predictive value of DDCHD of 74% (Table 1).

PGE_1 infusion was commenced in 50% (n = 71) of all infants, 63% of DDCHD group (n = 52), 18% of the PPHN group and all of the remaining non-DDCHD/non-PPHN group (Fig. 1). Median (range) PGE_1 dose was 20 (5–100) nanograms/kg/min for all infants.

Of the DDCHD group, 16 infants had duct-dependent pulmonary blood flow (PGE_1 used in 9); 8 pulmonary valve stenosis, 7 pulmonary atresia and 1 severe Ebstein's anomaly. Forty infants had duct-dependent systemic blood flow (PGE_1 used in 25); 27 coarctation, 4 severe aortic valve stenosis, 3 hypoplastic left heart syndromes and 6 hypoplastic/interrupted aortic arch. Twenty-five infants had mixing defects (PGE_1 used in 18); 16 transposition of the great arteries, 7 total anomalous pulmonary venous drainage, and 2 truncus ateriosus with aortic coarctation.

Demographic, clinical and transport characteristics are provided in Table 2 and discussed next. Comparison of PGE_1 versus no PGE_1 groups, and DDCHD PGE_1 versus DDCHD noPGE_1 groups was performed to assess factors that may have influenced the transport team's decision to commence PGE_1, as a proxy indicator of suspicion of DDCHD.

Demographic and Clinical Characteristics

All infants

Median (interquartile range) gestation was 39 (38–40) weeks, and birthweight was 3.3 (2.9–3.8) kg. The commonest clinical findings were heart murmur (56%), cardiomegaly on CXR (44%) and cyanosis (37%).

PGE_1 versus noPGE_1 groups

Gestation and birthweight were similar between groups. In univariate analysis, PGE_1 use was significantly associated with

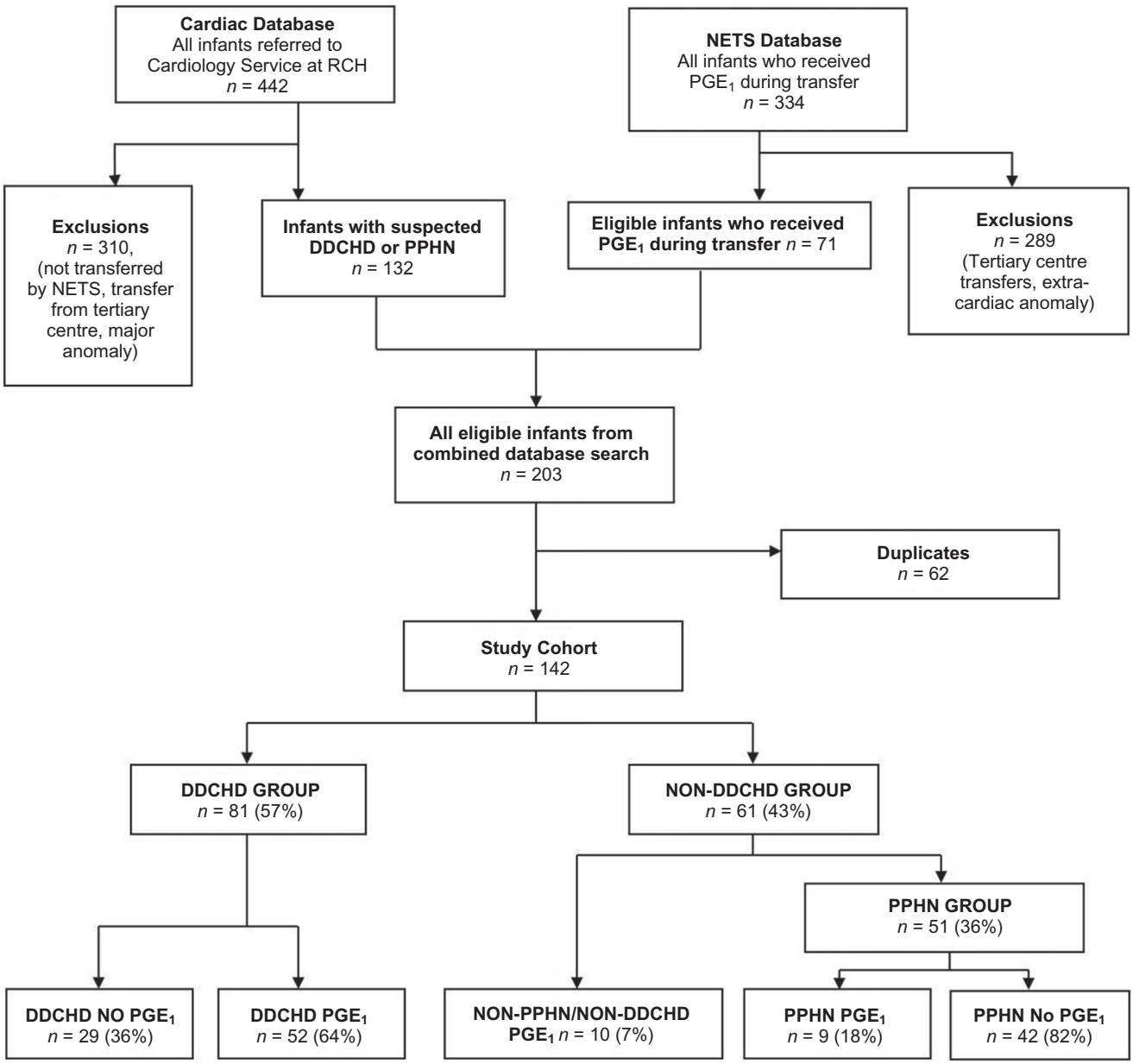

Fig. 1 Flowchart of identification of study population and subgroups from database analysis. DDCHD, duct-dependent congenital heart disease; NETS, Newborn Emergency Transport Service, Victoria; PGE_1, prostaglandin E_1; PPHN, persistent pulmonary hypertension of the newborn; RCH, Royal Children's Hospital, Melbourne.

the spontaneous vaginal delivery, Apgar score <5 at 5 min, cyanosis, heart murmur, abnormal pulses, and lower SpO_2 and PaO_2. On multivariate analysis, cyanosis and abnormal pulses were significantly associated with use of PGE_1.

DDCHD PGE_1 versus DDCHD noPGE_1

The median gestation and weight was similar in both subgroups. In univariate analysis, PGE_1 use was significantly associated with male sex and presence of heart murmur.

Table 1 Comparison of provisional diagnosis with echocardiography – confirmed diagnosis

Provisional diagnosis by transport team	Echocardiographic diagnosis	
	DDCHD (*n* = 81)	No DDCHD (*n* = 61)
Suspected DDCHD (*n* = 98)	73	25
No DDCHD (*n* = 44)	8	36

DDCHD, duct-dependent congenital heart disease.

Table 2 Demographic, clinical and transport characteristics of all infants

	All infants (*n* = 142)	PGE_1 group (*n* = 71)	Non-PGE_1 (*n* = 71)	*P*	DDCHD PGE_1 *N* = 52	DDCHD noPGE_1 *N* = 29	*P*
Demographic data							
Gestation, median (IQR)	39 (38–40)	39 (38–40)	39 (37–40)	0.12	39.5 (38–40)	39 (37–40.5)	0.11
Birthweight (g), median (IQR)	3.3 (2.9–3.8)	3.5 (2.9–3.7)	3.3 (2.9–3.8)	0.50	3.4 (3.0–3.7)	3.4 (2.5–3.8)	0.42
Male sex, *n* (%)	83 (58)	45 (63)	38 (53)	0.31	36 (69)	13 (45)	0.04
IUGR, *n* (%)	12 (8)	5 (7)	7 (9.8)	0.76	3 (6)	5 (17)	0.13
Mode of delivery (LSCS), *n* (%)	53 (37)	17 (24)	36 (50.7)	0.002	17 (33)	10 (34)	1.0
Clinical characteristics							
Apgar <5 at 5 min, *n* (%)	14 (10)	1 (1.5)	13 (18.3)	0.001	0 (0)	2 (7)	0.13
Cyanosis, *n* (%)	52 (37)	32 (45)	20 (28)	0.037**	22 (42)	13 (45)	0.83
Heart murmur, *n* (%)	80 (56)	46 (64.7)	34 (47.8)	0.042	36 (69)	26 (90)	0.04
Abnormal pulses, *n* (%)	35 (25)	27 (38)	8 (11.2)	0.0001**	23 (44)	8 (28)	0.14
Labile saturations, *n* (%)	40 (28)	18 (25)	32 (45)	0.014	6 (12)	1 (3)	0.21
Pre-post ductal difference, *n* (%)	46 (32)†	26 (39)†	20 (41.6)†	0.8	18 (37)†	6 (25)†	0.32
Upper and lower BP difference > 10, *n* (%)	30 (21)†	20 (32)†	10 (22.7)†	0.3	19 (41)†	8 (33)	0.52
PaO_2 at NETs arrival (mean, SD)	50 (20)†	47 (21)†	59 (22)†	0.04	48 (23)†	44 (14)†	0.97
SpO2 at NETS arrival (mean, SD)	85 (15)†	84 (16)†	88 (14)†	0.13	83 (17)†	90 (10)†	0.04
Mean BP at NETS arrival (mean,SD)	50 (14)†	54 (15)†	47 (13)†	0.005	55 (16)†	49 (14)†	0.16
Abnormal parenchyma (CXR), *n* (%)	32 (22)	8 (11.2)	24 (33.8)	0.01	3 (6)	3 (14)	0.45
Cardiomegaly (CXR), *n* (%)	63 (44)	41 (57.7)	22 (31)	0.02	34 (65)	19 (65)	0.99
Transport characteristics							
Distance (km), mean (range)	63 (21–150)	63 (21–140)	63 (26–152)	0.72	63 (30–128)	64 (24–178)	0.32
Land, *n* (%)	101 (71)	48 (68)	53 (75)	0.46	34 (65)	21 (72)	0.63
Air, *n* (%)	41 (29)	23 (32)	18 (25)	0.46	18 (35)	8 (28)	0.63
Time critical, *n* (%)	84 (59)	48 (68)	36 (51)	0.06	36 (70)	9 (31)	0.0012
Primary urgent, *n* (%)	56 (40)	22 (31)	34 (48)	0.06	15 (29)	19 (66)	0.0021
Non-urgent, *n* (%)	2 (1)	1 (1.5)	1 (1.5)	1.0	1 (2)	1 (3)	1.0
Total time of transport (h) mean (range)	5.4 (1–34)	5.9 (2–34)	4.9 (1–10)	0.05	5.4 (2–12)	4.4 (1–8)	0.04

***P* < 0.05 on multivariate analysis. †Missing data. BP, blood pressure; CXR, chest X-ray; DDCHD, duct-dependent congenital heart disease; IQR, interquartile range; IUGR, intrauterine growth retardation; LSCS, lower segment Caesarean section; NETS, Newborn Emergency Transport Service, Victoria; PGE_1, prostaglandin E_1.

Transport characteristics

Mean (range) transfer distance was 63 (21–150) km, and mean (range) transport duration was 5.4 (1–34) h. Seventy-one per cent of transfers were by land (29% by air), and 59% were classified as 'Time Critical'.

Factors distinguishing DDCHD and non-DDCHD

On univariate analysis, heart murmur, abnormal pulses, upper and lower limb BP difference >10 mmHg, cardiomegaly, initial SpO_2 of <92%, PaO_2 <50 mmHg, and pre-post ductal SpO2 difference >10% were significantly associated with DDCHD (Table 3). None of these clinical factors were significantly associated with DDCHD on multivariate analysis.

Labile SpO_2, abnormal lung parenchyma, mean BP < 40 mmHg, pH < 7.25, lactate >5 and FiO_2 ≥0.5 were significantly associated with non-DDCHD. On multivariate analysis, only mean BP ≤40 mmHg and labile SpO2 significantly associated with non-DDCHD, abnormal lung parenchyma approached significance ($P = 0.05$).

Effect of PGE_1 use on clinical status

Comparing physiological variables at time of NETS arrival at the referring centre (Time I) and arrival of NETS at the tertiary cardiac centre (Time II) (Table 4), FiO_2 was significantly lower in both PGE_1 and noPGE_1 groups after transfer. No other physiological variable was significantly altered in either group during the retrieval and transfer (i.e. between Times I and II). The effects of PGE_1 on cardiorespiratory status and outcomes in this cohort were the focus of a separate analysis and report. Rates of ventilation and inotrope use did not differ between PGE_1 and noPGE_1 groups.[7]

Discussion

Our study confirms that a large number of infants with suspected DDCDH are delivered without antenatal diagnosis, in non-tertiary settings, long distances from a paediatric cardiac centre. The resources mobilised to retrieve and transfer these infants are substantial. Making an accurate diagnosis early has important implications for therapy and transfer.

Table 3 Predictors of DDCHD

Variable	DDCHD	No DDCHD	Univariate	Multivariate analysis	
	N = 81	*N* = 61	*P*	*P*	AOR, 95% CI
Cyanosis	35/81	17/61	0.06	–	–
Heart murmur	62/81	18/61	**<0.001**	0.85	1.1, 0.3–4.1
Abnormal pulses	31/81	4/61	**<0.001**	0.33	2.1, 0.5–9.4
Labile saturations	7/81	43/61	**<0.001**	**0.005**	0.1, 0.0–0.5
Pre-post ductal saturation difference >10%	24/73	22/42	**0.04**	0.41	0.6, 0.2–2.0
CXR – abnormal parenchyma	6/81	26/61	**<0.001**	**0.05**	0.2, 0.1–1.0
CXR – cardiomegaly	53/81	10/60	**<0.001**	0.90	1.1, 0.3–4.7
BP upper and lower limit > 10 mmHg	27/70	3/36	**0.01**	0.33	2.2, 0.5–11.0
SpO_2 at referral <92%	28/43	9/24	**0.03**	0.52	1.9, 0.3–14.6
MBP at referral <40 mmHg	15/71	22/43	**0.001**	**0.03**	0.15, 0.03–0.4
Lactate at referral >5	6/32	17/26	**0.0001**	0.52	0.5, 0.1–4.40
PaO_2 at referral <50 mmHg	28/43	9/24	**0.03**	0.35	0.4, 0.1–3.0
pH at referral <7.25	21/51	35/41	**0.0001**	0.27	0.3, 0.04–2.43
BE at referral ≤10	11/49	21/40	**0.003**	0.53	2.2, 0.2–23.5
FiO_2 >0.5	32/81	33/61	0.08	–	–

Bold indicates significant *P* value. AOR, adjusted odd ratio; BE, base excess; BP, blood pressure; CXR, chest X-ray; DDCHD, duct-dependent congenital heart disease; MBP, mean blood pressure.

Table 4 Physiological parameters in PGE_1 and noPGE_1 groups during transport

Physiological parameter		All PGE_1 (*n* = 71)	All noPGE_1 (*n* = 71)	*P* (PGE_1 vs. noPGE_1)
SaO_2	Time I	84 (16.3)	88 (13.8)	0.13
Mean(SD)	Time II	87 (13.4)	92 (7.5)	0.008
	P*	0.5	0.5	
FiO_2	Time I	55.5 (33.4)	60.0 (33.5)	0.5
Mean (SD)	Time II	40.0 (26.7)	47.3 (28.3)	0.12
	P*	0.01	0.01	
Mean BP	Time I	53.8 (14.8)	47.1 (13.0)	0.005
Mean (SD)	Time II	50.6 (13.0)	46.0 (10.3)	0.021
	P*	0.34	0.91	
pH	Time I	7.18 (0.21), *n* = 44	7.18 (0.18), *n* = 32	0.98
Mean (SD)	Time II	7.23 (0.17), *n* = 41	7.23 (0.16), *n* = 42	0.99
	P*	0.5	0.25	
Lactate	Time I	5.9 (5.0), *n* = 33	6.0 (5.3), *n* = 24	0.93
Mean (SD)	Time II	6.0 (4.6), *n* = 28	5.9 (5.1), *n* = 37	0.96
	P*	0.44	0.65	

P*: comparison of Time I versus Time II. Time I: Arrival of NETS Team at referring centre. Time II: End of retrieval/arrival of NETs Team at tertiary cardiac centre. PGE_1, prostaglandin E_1.

Although diagnostic accuracy by NETS staff was relatively high, and comparable with previous reports,[8] nearly one quarter of infants with DDCHD did not receive PGE1. The decision to start PGE_1 was strongly associated with abnormal pulses, though also associated with evidence of hypoxia (cyanosis and low PaO_2), murmur and low Apgar score. Within the group of infants with DDCHD, the presence of a murmur appeared to be an important discriminator of PGE_1 use, too. This may suggest that the managing team considered these findings most likely to distinguish DDCHD from non-DDCHD. However, PGE_1 use may not be an entirely accurate proxy of suspected DDCHD. The transport team may have reserved PGE_1 use only for the 'sick suspected DDCHD' infants, and in a more 'stable suspected DDCHD' group may have elected not to commence PGE_1. Accordingly, the finding that only 64% of infants with DDCHD received PGE_1 may indicate rationale

patient selection, rather than poor clinical judgement. The stability of physiological parameters during transfer, irrespective of PGE_1 use, is evidence of appropriate clinical decision-making by the transport team.

Which clinical findings actually help distinguish DDCHD from non-DDCHD? Definitive diagnosis confirmed that heart murmur and abnormal pulses were indeed significantly associated with DDCHD, as were upper and lower limb BP difference, cardiomegaly, low SpO2 and pre-post ductal SpO2 difference. Cyanosis, however, was not, and none of these factors were significant on multivariate analysis.

Non-DDCHD was associated with parenchymal changes on CXR, acidosis and higher FiO_2, but on multivariate analysis only with labile SpO2 and hypotension (mean BP < 40 mmHg). Overall, non-DDCHD infants appeared to be more unwell, which may reflect a population of infants with PPHN who are more unstable than those with isolated DDCHD.

Our findings are in agreement with those of Danford *et al*. who identified abnormal pulses and murmur as important predictors of PGE_1 responsiveness.[9] In contrast to our findings, however, this group identified cyanosis as the strongest predictor of PGE_1 responsive cardiac lesions. In a Canadian cohort, Shivananda *et al*. found no association between clinical findings and echocardiographic diagnosis of CHD in hypoxaemic infants, though X-ray evidence of parenchymal lung disease, FiO_2 and SpO2 were significant discriminators.[8] Pickert *et al*. also observed that clinical findings were poor discriminators of CHD, when comparing obstructed left heart lesions and sepsis.[10] The same study highlighted the utility of cardiomegaly on CXR to identify left heart anomalies, observing a sensitivity of 85%, and positive predictive value of 95%.

The variation between these studies reflects the problems of discriminating DDCHD based on clinical findings alone. The addition of CXR and blood gas data may assist an informed judgement. Based on our own findings, we propose a stratification of combined findings to distinguish DDCHD from non-DDCHD (Table 4). Prospective assessment of accuracy of this guide is necessary before it can be recommended for use in clinical practice.

Accepting the limitations of clinical findings, which other methods could assist the diagnosis of DDCHD in this setting? Previous studies have shown that electrocardiogram (ECG) changes can be highly sensitive and specific in differentiating cardiac disease from PPHN.[11] Although an ECG may be readily performed at most referring centres, the skills in interpretation, to identify the changes associated with congenital cardiac disease, may not.[12]

Echocardiography at the referring centre would provide a definitive diagnosis. However, this requires an experienced echocardiographer and a suitable ultrasound device, neither of which may be available, even in a tertiary setting. Telemedicine may be a useful alternative;[13] If an ultrasound device and appropriate data connection are available, a referrer, with limited ultrasound experience, can demonstrate basic cardiac views to a remote paediatric cardiology team and a diagnosis may be made. Using telemedicine in suspected neonatal CHD, Mulholland *et al*., in Northern Ireland, reported a diagnostic accuracy of 93% and avoided transfer of infants in 74% of cases.[14]

A potential alternative, or adjunct, to telemedicine is the use of echocardiography by the retrieving team. Suitable portable ultrasound equipment is now available, and the development of ultrasound training aimed at neonatologists may be a means of providing transport staff with appropriate skills.[15] Further research in this area may be an important step towards improved diagnosis and management of infants with DDCHD during retrieval.

Historically, a principle reason for distinguishing DDCHD from non-DDCHD has been to ensure that PGE_1 is administered in DDCHD but to avoid its use in non-DDCHD, where it has been considered to confer no therapeutic advantage and expose to side effects including apnoea or hypotension.[5,16] However, recent studies provide reassurance that infants may be safely transported on PGE_1 particularly when lower doses are used. Browning Carmo *et al*. observed that within a group of infants with CHD receiving PGE_1, who were not electively intubated, 83% did not subsequently require respiratory support.[17] In a US cohort of infants receiving PGE_1 during transport, 36% of infants were transferred without intubation, and respiratory and cardiovascular complication rates were significantly lower compared with those who were electively intubated for transfer.[18]

We have additionally analysed the effects of PGE_1 use in our cohort in a separate recently published report.[7] PGE_1 use was not associated with increased use of invasive ventilation or inotropes, in either DDCHD or non-DDCHD, including PPHN. Furthermore, PGE_1 use was not associated with any deterioration in cardio-respiratory parameters during transfer. These data

Table 5 Proposed guide for distinguishing DDCHD and non-DDCHD

	'Predictive of DDCHD'	'Predictive of non-DDCHD/PPHN'
'Stronger predictors' (significant on multivariate analysis)		Mean BP < 40 mmHg (term infants) Labile SpO2
'Weaker predictors' (significant on univariate analysis only)	Heart murmur Abnormal pulses Cardiomegaly on chest X-ray Upper and lower limb BP difference >10 mmHg PaO_2 < 50 mmHg	Pre-Post ductal SpO2 difference >10% Abnormal lung parenchyma on chest X-ray Lactate >5 Low pH (<7.1) Low BE (<–8.0)

BE, base excess; BP, blood pressure; DDCHD, duct-dependent congenital heart disease; PPHN, persistent pulmonary hypertension of the newborn.

allay concerns regarding side effects of PGE_1. Based on these findings, and in light of the challenges of distinguishing DDCHD on clinical grounds alone, it remains prudent and safe to commence PGE_1 if DDCHD is suspected.

Limitations

Our study is limited by its retrospective nature; in some cases original documentation was incomplete, as noted.

Conclusions

Distinguishing DDCHD from PPHN remains a major challenge during the retrieval of hypoxic infants, even for experienced neonatal teams. We propose a stratification of combined clinical findings, CXRs, SpO_2 and blood gases which may assist discrimination of DDCHD and non-CHD (Table 5). In the absence of a reliable single clinical discriminator of DDCHD, PGE_1 should be used whenever DDCHD is suspected.

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