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Does Measurement of Four Limb Blood Pressures at Birth Improve Detection of Aortic Arch Anomalies?

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Abstract

Objective—To determine normal four-extremity blood pressure (BP) in the NICU at birth and the utility of upper (UE) and lower extremity (LE) BP difference to screen for coarctation of aorta (Co-A) and interrupted aortic arch (IAA).

Study Design—Retrospective study of BP at birth (n=866), and case-control study of Co-A/IAA infants and matched controls (1:2).

Results—Although BP increased with gestational age (R^2=0.3, p<0.0001), the pressure gradient between UE and LE did not change with gestation (p=0.68). Forty-six cases of Co-A/IAA were identified, with 92 controls. Pressure gradient was significantly higher in patients with Co-A/IAA (7.6±14.8 vs.0.4±10mmHg, p=0.004). However, there was overlap between cases and controls resulting in low sensitivity (41.3% with ≥10mmHg gradient cut-off).

Conclusion—Evaluation of UE-LE BP gradient at birth is a poor screening test for Co-A/IAA with low sensitivity. Repeating four-limb BP after ductal closure at 24–48h along with SpO_2 screening for critical congenital heart disease may increase sensitivity.

Keywords

Aortic arch anomalies; coarctation; interrupted arch; CCHD screening

INTRODUCTION

Congenital heart disease can present at different ages in an infant, depending on the type of defect. The implementation of critical congenital heart disease (CCHD) screening for all newborns in the USA has improved detection of certain cyanotic heart lesions. However, aortic anomalies were not intended to be detected by this screen[1] and currently there are no specific guidelines to screen for these conditions. Coarctation of the aorta (Co-A) is a narrowing of a segment of the aorta near the ligamentum arteriosum adjacent to the

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subclavian artery. It may be discrete, long or complex and may involve the aortic arch or isthmus. It accounts for 7% of liveborn children with congenital cardiac malformations, with a higher incidence in stillborn infants[2]. Although optimal age at intervention is not currently known[3], untreated Co-A has a high mortality rate - up to 80% by age 50[4]. Interruption of the aortic arch (IAA) accounts for over 1% of CCHD cases, with a known association between chromosome 22q11 deletion and DiGeorge Syndrome[2]. It results when there is regression of both the right and left fourth aortic arches during embryologic development. The site of regression can vary in each aortic arch resulting in different anatomic forms depending on the site of interruption[5]. Currently, surgical correction is attempted during the first week of life[6], hence timely recognition is critical for appropriate management of these infants.

Both conditions (Co-A and IAA) usually present around 2–7 days of age, at ductal closure, with circulatory collapse, shock, congestive heart failure and/or respiratory distress[7]. Normally, systolic pressure in the lower limbs in children may be up to 20mmHg higher than in the arm; a femoral systolic pressure lower than that in the arms is abnormal and suggests the need for further assessment[8]. Blood pressure measurements can serve as a useful and non-invasive tool for the diagnosis of Co-A in newborns. Measurement of four-extremity blood pressures at birth or admission is practiced in some newborn nurseries and neonatal intensive care units (NICUs) to look for an upper extremity (UE) to lower extremity (LE) pressure gradient to rule out Co-A and IAA. After ductal closure, blood pressures are lower in the lower extremities compared to upper extremities in Co-A and IAA[7]. However, there are no studies conducted to validate this protocol of checking blood pressures at birth, prior to ductal closure. We hypothesized that upper extremity blood pressures would be significantly higher than lower extremity blood pressures at birth in patients who are subsequently diagnosed with Co-A and IAA compared to gestational age matched controls.

METHODS

The Children and Youth IRB at University at Buffalo approved the study and waived the need for individual consent. Four-extremity blood pressures are routinely performed at birth, on admission to the NICU at Women and Children’s Hospital of Buffalo. An appropriately sized cuff is used to measure arm and leg blood pressures. Infants with an antenatal diagnosis of CHD were not included in this review. However, on retrospective chart review, two of the cases had poor visualization or suspicious antenatal sonogram and were recommended to have a postnatal echocardiogram by the obstetrician. The study consisted of two parts – a retrospective study of upper extremity to lower extremity pressure gradient and a case-control study within a population of infants admitted to the NICU.

a. Retrospective study of controls to detect normal blood pressure gradient at birth: Four-limb blood pressures from all babies admitted to the NICU from Sep 2011 through October 2012 were collected using the electronic medical record. This formed a pool of data from 866 newborns without the diagnoses of major cardiac defects (including aortic arch anomalies) detected either during prenatal period, during the NICU stay or for one-year after discharge from the NICU [all heart defects with the exception of patent ductus arteriosus (PDA), patent foramen ovale (PFO) and small < 5mm ventricular septal defects (VSD) were excluded].

Correlations between postmenstrual age (PMA) at birth and blood pressure and UE-LE pressure gradient were evaluated.

b. Case-control study: Study cases were obtained, comprising of all newborns discharged from the NICU from Jan 2000 through March 2014 with a diagnosis of aortic arch anomalies i.e. Co-A or IAA made either during the NICU stay or after discharge. Diagnoses of aortic arch anomalies was based on 2-D echocardiography results. Cases were matched for month and year of birth, gender and gestational age with 2 controls for each case. Patients in the control group had diagnoses other than aortic arch anomalies. A total of 46 patients in study group and 92 in control group met our criteria and their data was evaluated. Four limb systolic, diastolic, and mean blood pressures for all these newborns taken at the time of admission was recorded using appropriate sized cuffs. In both the groups, the difference in systolic, diastolic, and mean blood pressures between right upper extremity (RA) and right lower extremity (RL) and between left UE (LA) and left LE (LL) was recorded. Data on heart rate and pulse oximetry on admission, along with blood gases, were all recorded.

c. Statistical analysis: Pearson's correlation coefficient was used to compare blood pressure values and gestation. Given the almost normal distribution for all variables, student t-test was used to compare the blood pressure values, and other variables between cases and controls. Non-parametric tests were used for skewed data. Receiver-operating characteristic (ROC) curves were generated using SPSS 22 software (IBM Corporation, Armonk NY).

d. Sample-size calculation: We planned a study of a continuous response variable (difference between upper limb and lower limb pressure) from controls and cases of aortic arch anomalies with 2 controls per case. The highest difference between the upper extremity mean blood pressure and lower extremity mean blood pressure from 866 NICU admissions was normally distributed with a median of 0 and a standard deviation of 7.8 mmHg. If the true difference in the case and control means is 5 mmHg, we needed to study 39 experimental subjects and 78 control subjects to be able to reject the null hypothesis that the population means of the experimental and control groups are equal with probability (power) 0.9. The Type I error probability associated with this test of this null hypothesis is 0.05. With the current inclusion of 46 cases and 92 controls, we achieved a power of 0.942. [calculated with the Power and Sample size calculator from Vanderbilt University http://biostat.mc.vanderbilt.edu/wiki/Main/PowerSampleSize)]

RESULTS

Retrospective study

In the 866 newborns studied, systolic, diastolic and mean blood pressures correlated with increasing gestational age ($p<0.0001$). Figure 1 shows a scatter diagram with mean blood pressure in right arm and right leg plotted against postmenstrual age at birth. As expected, with increasing gestational age at birth, an increase in the mean BP in the arm and calf were observed. However, the differences between arm and leg blood pressures did not change with gestational age (Figure 2, p=0.68).

Case control study

We identified 31 cases of Co-A and 15 cases of IAA. All but one of these infants were diagnosed prior to discharge from the NICU. This one patient, after discharge, was noted to have absent femoral pulses during a routine 2-week check-up and was diagnosed with IAA. The following characteristics were compared between cases and controls - gestational age, birth weight, systolic, diastolic and mean blood pressures in all four limbs at birth, cord gas, heart rate and pulse oximetry on admission between the two groups (Table 1). Except for systolic pressure comparison in right upper and right lower extremity, we found a statistically significant difference between upper extremity and lower extremity blood pressures (systolic, diastolic and mean) in cases compared to controls (Table 1) regardless of statistical test performed. We noted that pulse oximetry readings (site – pre vs. post-ductal, not specified in the medical record) on admission were lower in cases than controls.

Among the cases of Co-A/IAA, 21/46 (45.6%) were noted to have decreased femoral pulses at the time of diagnosis. Five out of 46 (10.8%) patients were noted to have good femoral pulses. In the remaining cases, femoral pulse examination results were missing in the medical record. Table 2 outlines presenting clinical features in patients with aortic arch abnormalities. Shock, decreased perfusion and/or metabolic acidosis was the most common presentation.

Additionally, we looked at the values of blood pressure gradient between upper and lower limbs in the entire cohort of normal newborns (n=866) and those under study (controls n=92, cases n=46). In the whole cohort of 866 infants, the absolute difference was normally distributed with 30% of patients recording an UE-LE pressure gradient of 1 to 5mmHg (Figure. 3). In the case-control study, the controls demonstrated a similar distribution of the pressure gradient between upper and lower extremities with 30% of patients recording a gradient of 1–5 mmHg (Figure 4). In the study cases (patients subsequently diagnosed with Co-A/IAA), the upper limb pressures were significantly higher in cases than controls (Table 1 and Figure 4). The mean arm BP was greater than the calf BP in 65% of controls and 76% of cases (p: not significant).

We ascertained the cut-off value for difference between UE and LE blood pressures in the patients with aortic arch anomalies. We tested the sensitivity and specificity of mean blood pressure difference of 5, 10, and 15 mmHg. At a 15 mmHg cut-off, the specificity was 93.48%. As the cut-off increased, specificity increased with fewer false positives (Table 3). All the cut-off values yielded low sensitivity and the area under ROC curves area was low (systolic BP – 0.580, diastolic – 0.615 and mean – 0.626, curves not shown).

DISCUSSION

Four-extremity cuff measurements of BP are routinely obtained at birth by some neonatal intensive care units including ours, without much being known about the utility nor the interpretation of randomly timed measurements. Although there is known association of UE and LE pressure differences of up to 18mmHg in older children (4–18 years of age) with coarctation[8], there are currently no guidelines as to the optimal age of cuff pressure measurement after birth to assess for these conditions. To our knowledge, this is the first study attempting to validate the practice of obtaining four-extremity BP in all newborns admitted to a NICU at birth.

In the current study, blood pressure values were obtained shortly after birth, as our institution measures four limb blood pressures on admission, regardless of gestational age. The absolute increase in mean pressures with gestational age is a known trend[9], and substantiated by our results (Figure 1). Although indwelling arterial catheter measurements are the method of choice for accurate pressure monitoring[10], non-invasive oscillometric cuff pressures have been shown to correlate closely with indwelling arterial catheters, although cuff pressures may over-estimate values[11] and are especially inaccurate in extremely-low-birth-weight infants. However, the only reasonable way of doing the upper/lower blood pressure comparison is with non-invasive methods, but in small preterm babies they are very inaccurate. Given the consistency of mean pressure difference between UE and LE at birth despite a range of postmenstrual age at admission (Figure 2), we hypothesized that four-extremity cuff pressures will be effective in detecting potential aortic arch anomalies at all gestational ages.

Consistent with our hypothesis, we found a statistically significant increase in UE-LE pressure gradient at birth in cases who were subsequently diagnosed to have Co-A and IAA. We found a very variable largest mean UE-LE blood pressure difference in infants without cardiac anomalies, with a mode value of 1 to 5 mmHg gradient, and a range between more than +15 to below −15 mmHg (n=866, Figure 3). However, there was considerable overlap in the pressure gradient among study cases and controls. It is interesting to note that about a quarter of our study cases did have normal mean blood pressure difference, with a small proportion even having LE pressures up to 15mmHg higher than UE (Figure 4). While such high pressure gradients are known to occur in older children, they are uncommon in neonates[8]. Because of the high overlap in UE-LE pressure gradient at birth between cases and controls, this difference although statistically significant, may have poor clinical relevance.

On secondary analysis of right and left UE blood pressures among the 46 cases, 14 patients (30%) had a >10mmgHg difference between any UE and any LE. It was noted in our data, simply comparing the RA and any LE, only one case (with a large PDA on echocardiogram) would have been missed. This suggests that measuring pressures of the only right upper and any lower limb would save time and offer better efficiency with similar detection rates.

Cases of Co-A with normal blood pressures have been previously identified[13], so it's essential to account for a thorough history and physical exam (including careful auscultation and evaluation of dysmorphic features) to increase suspicion for aortic arch anomalies and not rely solely on mean blood pressure differences. Only two arch anomalies out of 46 cases were prenatally suspected to have abnormal views on fetal echocardiogram. Other studies have reported similar low detection rates for aortic arch anomalies. A retrospective study of IAA from Children's Hospital of Boston reported that over a 21 year period, the prenatal diagnosis was achieved in 24% of IAA cases.[14] The detection rates for Co-A are much lower with a recent article from the Swedish registry reporting that only 3/90 cases were diagnosed antenatally. In this series, of the 46 infants who were discharged home after birth undiagnosed, 1 patient died at home and was diagnosed postmortem and 22 presented in circulatory failure at readmission or shortly thereafter[15]. Low antenatal detection rates combined with high morbidity and mortality associated with late diagnosis and excellent long-term prognosis with timely diagnosis emphasize the need for an early postnatal screen for aortic arch anomalies[15].

In our study, 21/26 (81%) of Co-A/IAA patients with documentation of femoral pulses and perfusion showed signs of lower limb hypoperfusion and/or shock (Table 2). Interestingly, out of 866 infants in the retrospective study, 26(3%) presented with hypotension, none of whom were identified cases of Co-A nor IAA. These patients required a fluid bolus and/or dopamine infusion within 12 hours of admission. Only one patient with diaphragmatic hernia born at 29 weeks gestation in a community hospital was transferred in a moribund state with shock. Three patients had pulmonary hypertension with systemic hypotension and 16 were extremely preterm infants (23–25 weeks gestation) with transitional hypotension. 9/46 patients had dysmorphic features or other major anomalies (Table 2). None of the controls had dysmorphic features consistent with a genetic syndrome.

Pulse oximetry difference was significant between control and cases. Although the CCHD screen was not intended to detect for these conditions[16], given this finding in our study, it is likely that some of the affected infants might have failed their CCHD screen, had they been screened by the second day of life. However, the detection rate for Co-A using pulse oximetry-based CCHD screen is low[17]. Lannering et al reported that only 4 out of 19 infants (21%) were detected by routine pulse oximetry screening due to low oxygen saturation values in the lower limb[15].

Limitations of our study were intrinsic to the time of the cuff measurements which were within hours of birth, as currently done in our institution. However, as we know, most aortic arch anomalies usually manifest by day 2–7 of life after ductal closure. We reviewed all infants admitted and transferred to cardiothoracic service with a diagnosis of aortic arch anomalies. We could have missed late presentations (beyond 28 days) among NICU graduates. We could have also missed infants discharged from the newborn nursery but these infants were not part of our study.

Arm and calf pressures are known to have intrinsic variability. Aside from variations seen with crying or moving infants, age in older infants also been shown to correlate better with calf pressure than arm pressure, possibly attributed to the motion of arms more frequently than legs[18] Repetitive measurement tends to be an approach used for older children[19] but this is not feasible in neonates so one four-extremity BP measurement is often used in assessment. Diastolic cuff pressure can vary between thigh and calf pressure[19] so it's imperative to compare arm pressure specifically to calf pressure as the same cuff can be used for both.

As previously described by Crossland et al[20], differences between UE and LE mean pressures may also be attributed to random variability. There were a considerable number of normal infants without arch anomalies in our study who exhibited a 10–15mmHg difference in mean pressures similar to that reported in a recent study[21]. This overlap between normal controls and study patients with diagnosed aortic arch anomalies who have a similar difference in UE and LE pressures reduced the sensitivity of oscillometric pressure gradient. Another potential cause of decreased sensitivity would be because of cuff size used for arm and calf measurements. The difference in size between the calf and upper arm will result in falsely raised LE BP if the same size cuff is used for both[20], thus reducing the positive predictive value of cuff measurements. Systolic pressure in the LE may be higher than UE because of peripheral BP amplification[22]. Accounting for all these variables in measurements, based on our data, an UE mean pressure difference more than 15mmHg than LE appears to give the best specificity but low sensitivity (Table 3) making this a poor screening test to detect an aortic arch anomaly[21].

CONCLUSION

Patients with aortic arch anomalies have higher pressure gradient between upper and lower extremities at birth. However, the clinical utility of measuring four-extremity BP at birth to detect Co-A or IAA is low[22]. Co-A and IAA were not intended to be detected and are commonly missed by CCHD screening, so ultimately, blood pressure measurements and physical exams are relied upon for diagnosis. Oscillometric cuff pressure, best measured after the first 24 hours of life or after ductal closure, may be useful to detect aortic arch anomalies and improve intervention prior to clinical deterioration. The addition of four-limb mean blood pressure measurement to CCHD screening on or after the second day of life may improve sensitivity and specificity for aortic arch anomalies. Further studies are necessary to assess the utility of four-extremity BP at 24–48h after birth to detect aortic arch anomalies and are currently being performed in our NICU.

Acknowledgments

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Abbreviations

Co-A	Coarctation of aorta
IAA	Interrupted aortic arch
NICU	Neonatal intensive care unit
AAP	American Academy of Pediatrics
BP	blood pressure
UE	upper extremity
LE	lower extremity

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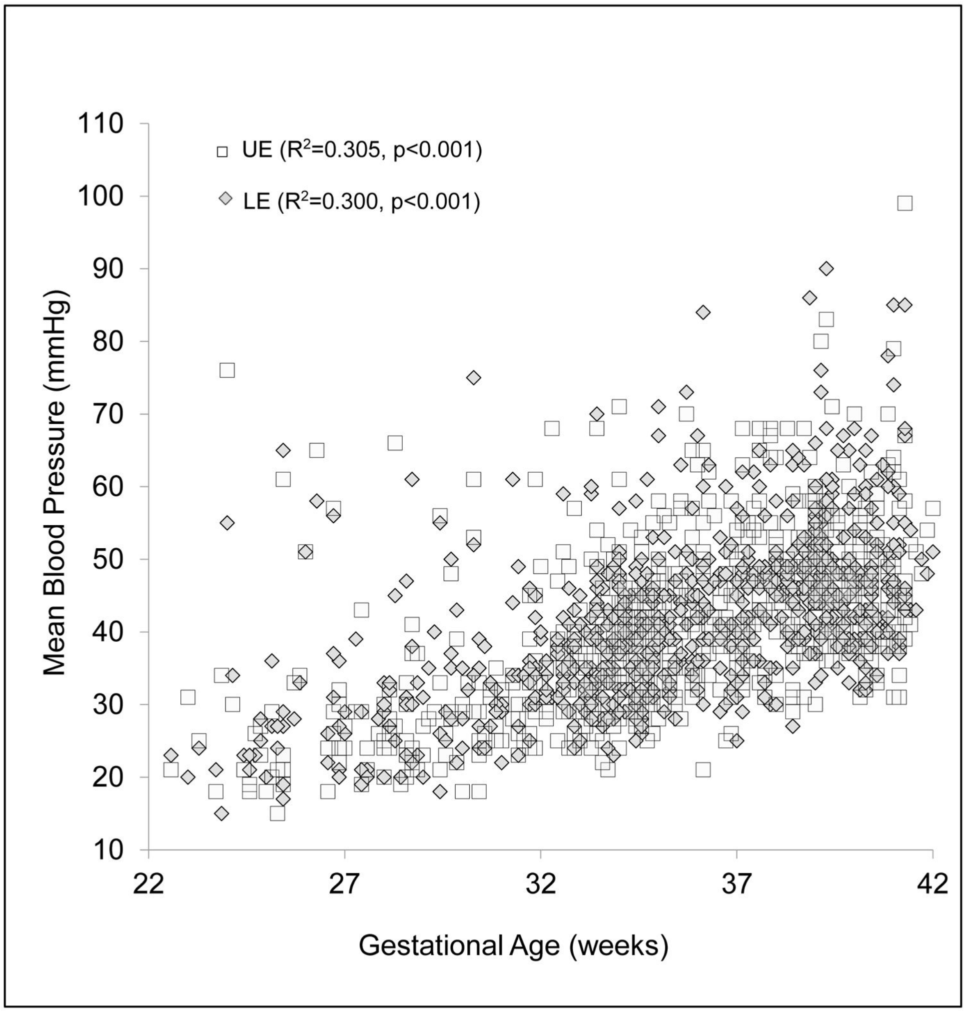

Figure 1.

Trend of upper extremity and lower extremity blood pressures at birth over increasing gestational age.

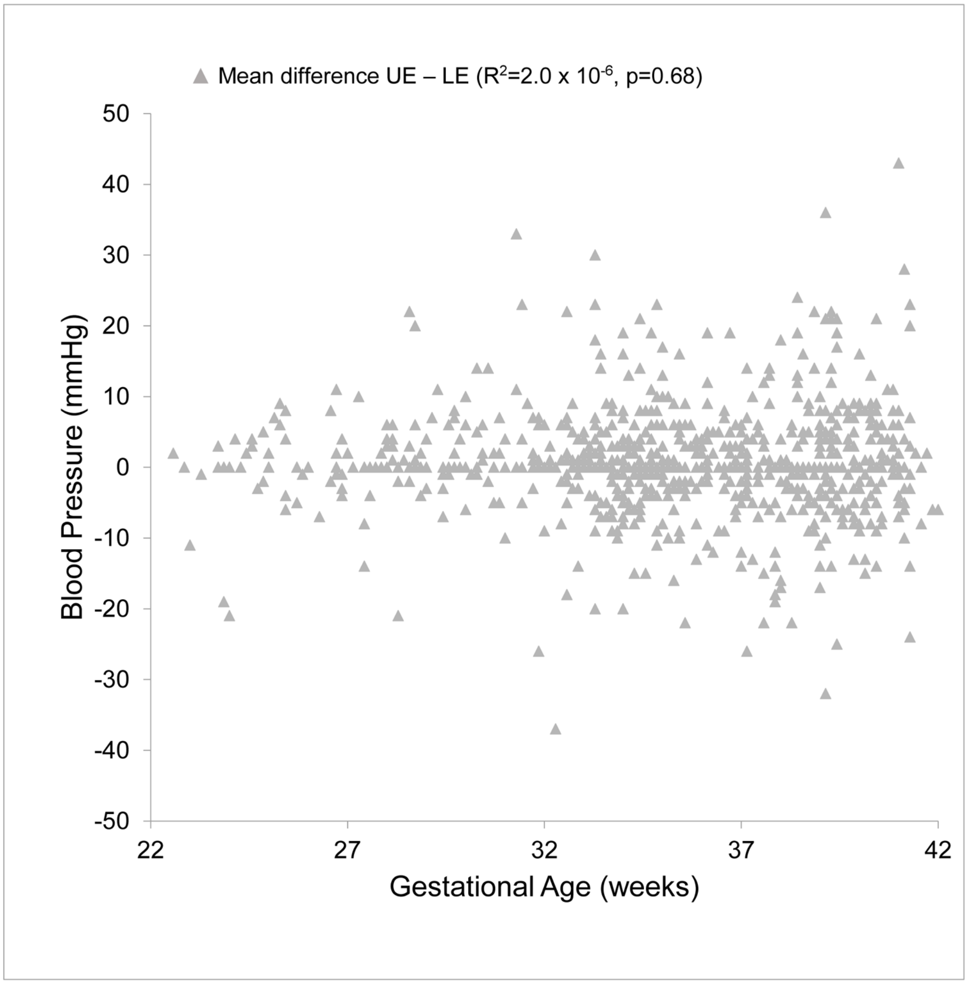

Figure 2.

Difference in mean blood pressure between upper extremity and lower extremity against gestational age at birth in all subjects (n=866).

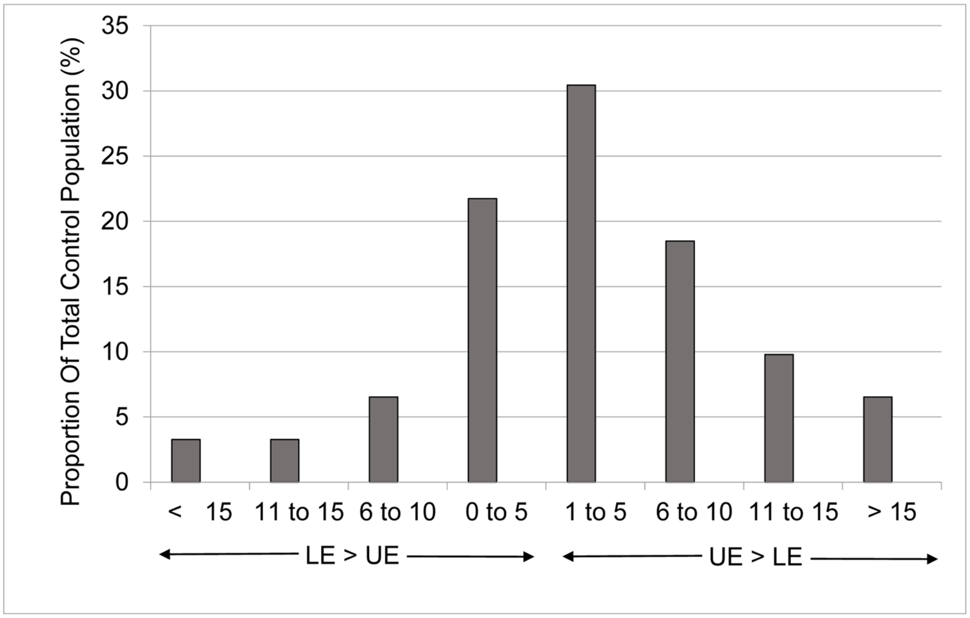

Figure 3.
Largest mean blood pressure difference between upper and lower limbs among all NICU admissions without major cardiac anomalies (n=866).

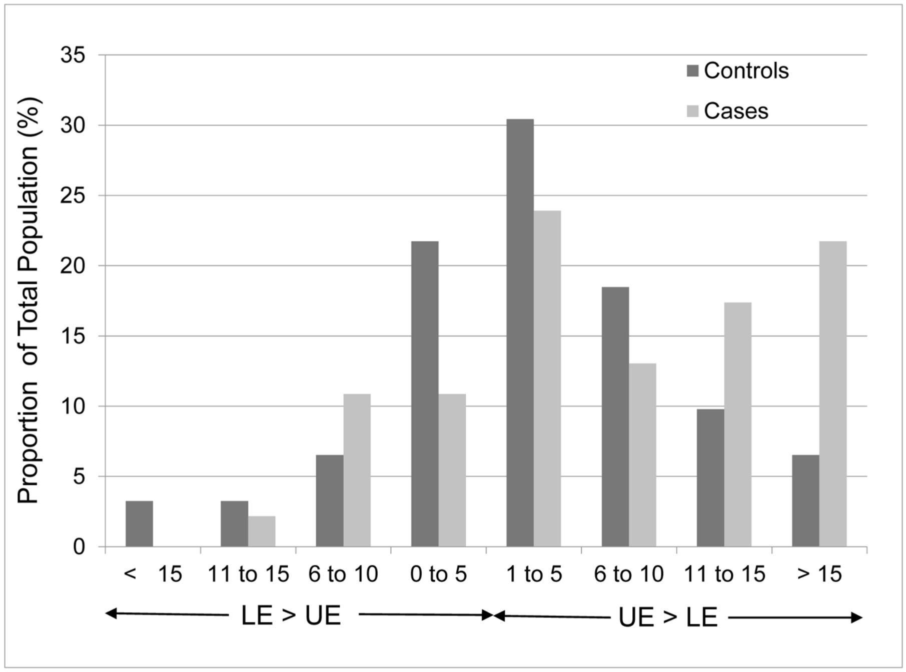

Figure 4.

Largest mean blood pressure differences between upper and lower limbs in cases of Co-A and IAA and matched controls.

Table 1

Demographic and blood pressure characteristics, SpO_2 and heart rate in the cases and the control groups.

Variables	Cases: Co-A, or IAA (mean ± SD) n=46	Control Group (mean ± SD) n=92	T-test P value	Non-parametric (Mann-Whitney U) test P value
Gestational age (weeks)	36.7 ± 3.5	36.7 ± 3.47	1	0.94
Birth weight (kg)	2.6 ± 0.9	2.62 ± 0.7	0.97	0.40
Right arm – Right leg systolic (mmHg)	7.2 ± 15.8	2.0 ± 9.8	0.03	0.92
Right arm – Right leg diastolic (mmHg)	6.9 ± 15.2	0.4 ± 10.7	0.01	<0.001
Right arm – Right leg mean (mmHg)	7.6 ± 14.8	0.4 ± 10.0	0.004	0.001
Left arm – left leg systolic (mmHg)	6.8 ± 16.9	1.2 ± 8.6	0.03	0.024
Left arm – left leg diastolic (mmHg)	6.6 ± 13.6	1.3 ± 7.9	0.01	0.005
Left arm – left leg mean (mmHg)	7.0 ± 13.5	1.3 ± 6.8	0.007	0.007
Pulse oximeter (%) Median (IQR)	96 (93–99)	98 (96–100)	0.002	0.009
Heart rate (beats/min)	146.5 ± 16.8	146.1± 16.9	0.88	0.864
Cord gas				
pH	7.35 ± 0.09	7.25 ± 0.76	0.35	0.27
Base Excess	−3 ± 3.1	−2.9 ± 4.8	0.90	0.15

Table 2

Presenting clinical features in patients with aortic arch abnormalities (some patients presented with more than one finding).

Clinical feature	Number of patients	Comments
Shock/ circulatory collapse/ metabolic acidosis / lower limb hypoperfusion	21	Two deaths and all survivors required neonatal surgical correction
Cyanosis / desaturations	8	7 – surgery in the neonatal period 1 – cardiology follow-up
Respiratory distress	7	1 – laryngeal web 6 – neonatal surgical correction 1 – cardiology follow-up
Hypertension (upper extremity)	1	Atenolol followed by surgery
Bloody stools (NEC)	1	Died
Hypoglycemia as an indication for NICU admission	4	Physical exam in the NICU concerning leading to echocardiogram
Dysmorphology – echocardiogram as part of workup	9	Anal fistula, absent radius -1 Imperforate anus - 1 Diaphragmatic hernia – 1 (died) Trisomy 21 -1 (no surgery required) Turner syndrome - 1 Williams syndrome - 1 Noonan syndrome –1 Apert syndrome – 1 (discharged home without surgery at 2 months) Di George syndrome - 1
Murmur only	3	2 – surgery (one after the neonatal period) 1 – follow-up by cardiology

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Table 3

Sensitivity and specificity of various cut-offs between upper extremity and lower extremity mean pressure gradient. The gold standard is detection of Co-A or IAA by echocardiogram.

Pressure gradient	Sensitivity	Specificity
≥ 5 mmHg	56.5%	59.8%
≥ 10 mmHg	41.3%	81.5%
≥ 15 mmHg	21.7%	93.5%