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Original Article

Cite this article: Thai L and Rochelson E (2025) Significance of northwest axis on neonatal electrocardiograms. *Cardiology in the Young* 35: 60–63. doi: [10.1017/S1047951124026465](https://doi.org/10.1017/S1047951124026465)

Received: 21 July 2024
Revised: 27 August 2024
Accepted: 29 August 2024
First published online: 24 January 2025

Keywords:

Northwest axis; CHD; neonatal electrocardiograms

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Abstract

Purpose: Northwest axis on an electrocardiogram is conventionally thought to be associated with CHD, but there is a paucity of data regarding the types and incidence of CHD associated with this finding. The purpose of this study was to report the types and incidence of CHD found at our institution to determine the efficacy of electrocardiograms as a screening test in infants. **Methods:** Retrospective, single-centre study of all infants ≤ 60 days of age who underwent a first-time electrocardiogram from 2015 to 2021. Our inclusion criteria included those found to have a northwest axis on electrocardiogram and who also had an echocardiogram performed. Data were described using standard summary statistics. **Results:** Two hundred sixty-eight infants met inclusion criteria (median age 4 days, **interquartile range** 2–24). The most common reason for an electrocardiogram to be obtained was the presence of a heart murmur ($n = 102$, 38%). One hundred thirty-one infants (49%) were found to have significant CHD. Cardiac defects included ventricular septal defect ($n = 36$, 28%), severe pulmonary valve stenosis ($n = 17$, 13%), atrioventricular canal defect ($n = 16$, 12%), and single ventricle disease ($n = 16$, 12%). **Conclusion:** In this study, nearly half (49%) of these infants with northwest axis on electrocardiogram were found to have significant CHD. Electrocardiograms remain an important screening tool for infants with findings associated with potential cardiac disease and echocardiogram is warranted for infants found to have northwest axis on a first-time electrocardiogram.

Introduction

CHD in the first few months of life can present with abnormal physical exam findings, such as cardiac murmurs or cyanosis.¹ Electrocardiograms are often used as a screening test to determine if further cardiac evaluation is needed for these infants. Evaluating the QRS cardiac axis on an electrocardiogram is an important factor to consider if there is an underlying CHD.

The QRS cardiac axis represents the sum of all the individual vectors generated by depolarisation waves and the net direction of the electrical current as it travels through the ventricular myocardium.^{2,3} It is important to understand the normal QRS axis in infants, as these values change over the first few months and years of life. These changes, in a structurally normal heart, are largely due to the physiologic shift from the dominance of the right ventricle in fetal life to the left ventricle in the more mature heart.³

A normal QRS for a newborn is approximately $+90^\circ$ to $+180^\circ$ and can be considered normal until about 6 months of age.^{2–4} Northwest axis, defined as a QRS axis of $+180^\circ$ to $+270^\circ$ (Figure 1), is conventionally thought to be associated with CHD.^{5,6} However, there is a paucity of data regarding the types and incidence of CHD that may be associated with this finding. The purpose of this study is to evaluate and describe the type and incidence of CHD among infants with northwest axis on electrocardiogram who also received an echocardiogram.

Methods

A single-centre, retrospective chart review of infants who had a first-time electrocardiogram obtained between January 1, 2015 and December 31, 2021 at The Children's Hospital at Montefiore (Bronx, NY) was performed. Infants who were ≤ 60 days of age, had a northwest axis (QRS axis $+180^\circ$ to $+270^\circ$) on electrocardiogram, and had an echocardiogram performed were included in our study. Patients who were older than 60 days of age, who did not have northwest axis on electrocardiogram, or did not have an echocardiogram to confirm if there were congenital structural abnormalities, were excluded. Patients were also excluded if the electrocardiogram was done incorrectly (for example, with limb-lead reversal). This study was approved by the Institutional Review Board of the Albert Einstein College of Medicine.

The decision to obtain a standard 12-lead electrocardiogram was determined by the clinical provider at the time of patient care. The reported QRS values were calculated automatically by clinical electrocardiogram software (GE MUSE™ Cardiology Information System, GE Healthcare) and confirmed by a paediatric cardiologist. The decision to obtain a transthoracic

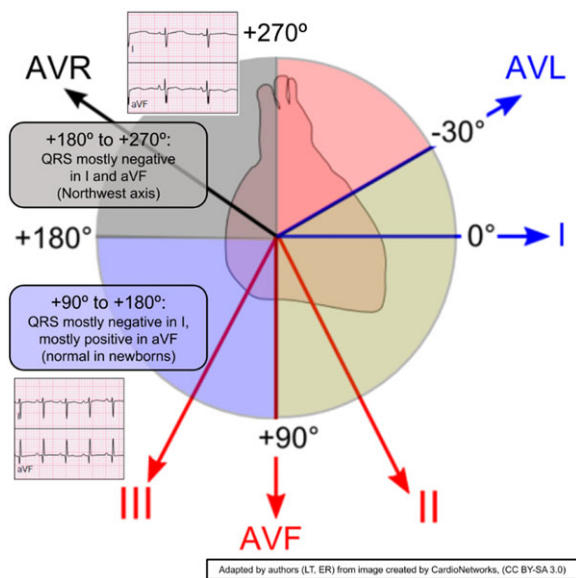


Figure 1. Frontal plane QRS axis.

echocardiogram was also at the discretion of the caring provider, as an echocardiogram was not always performed in all neonates with northwest axis at our institution.

For the patients who met our inclusion criteria, data regarding age, sex, indication for electrocardiogram, and echocardiographic findings for each patient were obtained. Significant CHD was defined as a congenital cardiac lesion that required close cardiac outpatient surveillance and/or required surgical intervention. As the aim of this study was to evaluate the value of electrocardiograms in screening for children with significant CHD, hemodynamically insignificant lesions such as a patent ductus arteriosus, small atrial septal defects, and small ventricular septal defects were excluded. These lesions typically do not require surveillance or intervention and could reasonably be discharged from cardiac care.¹⁸ Data were described using standard summary statistics.

Results

There were 2,276 infants ≤ 60 days of age who received an electrocardiogram during our study period at The Children’s Hospital at Montefiore. Out of those patients, a total of 325 infants were found to have **northwest axis** on their initial **electrocardiogram**. Out of 325 infants with **northwest axis**, 57 (17.5%) infants did not receive an echocardiogram, and therefore did not meet our inclusion criteria. A total of 268 infants (168 male, 100 female) met our inclusion criteria of having **northwest axis** on initial **electrocardiogram** and having had an echocardiogram performed.

In our study cohort, the median age was 4 days (interquartile range 1-24) and the median frontal plane QRS cardiac axis was 201° (IQR 189-223). The most common reason for an **electrocardiogram** to be obtained was the presence of a heart murmur ($n = 102$, 38%), but also included abnormal heart rates ($n = 33$, 12%), suspected syndrome ($n = 31$, 12%), known fetal diagnosis ($n = 29$, 11%), desaturations ($n = 25$, 9%), other (non-cardiac) reasons ($n = 28$, 10%), and a family history of CHD ($n = 4$, 1%, Table 1). The other, non-cardiac reasons for obtaining an **electrocardiogram** included patients undergoing a sepsis

Table 1. Patient data

Study population, n = 268	
Age in days	4 (2–24)
Male	168 (63%)
Frontal Plane QRS Axis (degrees)	201 (189–223)
Reason for Electrocardiogram	
Murmur	102 (38%)
Abnormal heart rates	33 (12%)
Suspected syndromes	31 (12%)
Fetal diagnosis	29 (11%)
Other	28 (10%)
Desaturations	25 (9%)
Family history	4 (1%)
Undocumented	16 (6%)

Note. IQR = interquartile range. The “other” category included reasons such as hypotension, electrolyte abnormalities, sepsis, seizures, cardiomegaly seen on chest radiograph.

evaluation, hypotension, electrolyte abnormalities, seizures, or cardiomegaly found on chest radiograph.

Of the 268 infants who met the inclusion criteria for our study, 131 (49%) were found to have significant CHD on echocardiogram. CHD types included large ventricular septal defect ($n = 36$, 28%), severe pulmonary valve stenosis ($n = 17$, 13%), atrioventricular canal defect ($n = 16$, 12%), and single ventricle disease ($n = 16$, 12%). Other defects included miscellaneous diagnoses, such as large coronary fistula to the right ventricle ($n = 5$), cardiac rhabdomyomas ($n = 4$), vascular ring ($n = 2$), coarctation of the aorta ($n = 2$), and dextrocardia ($n = 4$). Further delineation of the types of CHDs are listed in Table 2.

Discussion

There is a paucity of data evaluating the types and incidence of paediatric CHD associated with a northwest axis (+180° to +270°) on neonatal electrocardiograms. In this study, nearly half (49%) of infants ≤ 60 days of age who had northwest axis on electrocardiogram and had an echocardiogram were found to have significant CHD. To our knowledge, this is the largest study evaluating northwest axis in the neonatal period that describes the CHDs associated with this electrocardiographic finding.

Northwest axis deviation is a rare phenomenon, with an incidence estimated to be about 4%.⁶ Various case reports and studies have shown associations of northwest axis with CHD, ranging from transposition of the great arteries, single ventricle patients, Eisenmenger syndrome, Tetralogy of Fallot, severe pulmonary valve stenosis, and dextrocardia.^{6–8}

In addition to the aforementioned forms of structural heart disease that can cause northwest axis, other causes include inherited arrhythmia syndromes and conduction abnormalities. For example, some patients with Brugada syndrome have northwest axis on electrocardiogram, which is thought to be due to a pronounced conduction delay in the right ventricular outflow tract.⁸ Similarly, patients with bundle branch blocks and/or fascicular blocks may also demonstrate northwest axis.^{8,13,16} Tachyarrhythmias may cause northwest axis due to ventricular

Table 2. Study results

Presence of Congenital Heart Disease n (%)	
Patient without CHD	137 (51%)
Patients with CHD	131 (49%)
Types of Congenital Heart Disease	Patients with CHD Subset n = 131
VSD	36 (28%)
Severe PS	17 (13%)
AVCD	16 (12%)
Single Ventricle	16 (12%)
Moderate-to-Large ASD	8 (6%)
TAPVR	7 (5%)
Shone's Complex	6 (5%)
TOF	4 (3%)
Ebstein's Anomaly	3 (2%)
TGA	1 (1%)
Other	17 (13%)

Note. CHD = congenital heart disease, VSD = ventricular septal defect, PS = pulmonary valve stenosis, AVCD = Atrioventricular canal defect, ASD = atrial septal defect, TAPVR = total anomalous pulmonary venous return, TOF = tetralogy of Fallot, TGA = transposition of the great arteries. The "other" category includes miscellaneous diagnoses, such as large coronary fistulas to the right ventricle, cardiac rhabdomyomas, vascular ring, coarctation of the aorta, and dextrocardia.

tachycardia or aberrant His-Purkinje fibre conduction. Patients with twin AV nodes, such as in certain forms of heterotaxy, may have an unusual and variable QRS axis. Of note, perhaps the most common cause of northwest axis on electrocardiogram is limb-lead reversal, in which the right arm and leg electrodes are inadvertently placed on incorrect sides. This error must be identified to avoid misdiagnosis of northwest axis.^{14–15}

An electrocardiogram is often an initial screening test that is obtained in neonates with certain presenting symptoms, such as a murmur or cyanosis. Determining the cardiac axis on an electrocardiogram is an important step in determining the need for further work-up. The aetiology for northwest axis is not completely understood and may be different for each individual patient. The cause of northwest axis may be secondary to atrioventricular nodal displacement, such as in patients with complete atrioventricular canal defects and large ventricular septal defects, which causes anatomic variation of the bundle branches.^{1,13} Northwest axis may be due to right-sided structural anomalies, such as in patients with Tetralogy of Fallot, or severe pulmonary stenosis, which may cause right ventricular hypertrophy, right ventricular remodelling, and/or altered activation sequence in the right ventricle.^{5–7,16} Northwest axis may also be caused by cardiac malpositioning, such as in patients with dextrocardia.

The limitations of this study include its retrospective, single-centre nature, and relatively small sample size. Of note, we included patients with a known fetal diagnosis of CHD in our study population; this may be considered a limitation, as an electrocardiogram is not a true screening test when the diagnosis is already known. Patients with a known fetal diagnosis of CHD only constituted 11% ($n = 29$) of our study population. However, we accept this limitation as part of our study, as excluding these

patients may lead to an underestimation of the association between northwest axis on electrocardiogram and CHD. Lastly, our study focused specifically on infants with northwest axis to then determine if CHD was present. We did not analyse all infants who had an electrocardiogram obtained without northwest axis, as these infants were less likely to have received an echocardiogram to determine if CHD was present. Therefore, we cannot accurately determine sensitivity or specificity. In addition, we report the incidence of CHD in patients of northwest axis; however, we cannot comment on the incidence of northwest axis in this general patient population.

Conclusion

An electrocardiogram remains an important screening tool for infants with signs and symptoms that may suggest CHD, such as murmur or oxygen desaturation. Our study supports that clinical providers should consider obtaining an echocardiogram in neonates who have signs and symptoms of possible CHD and are found to have northwest axis on their electrocardiogram.

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