Heart Murmurs in the Neonate

**An approach to the neonate with a heart murmur**

This guideline is applicable to medical and nursing staff caring for neonates in the West of Scotland.

**Introduction**

- A heart murmur heard in the neonatal period may be associated with congenital heart disease.

- However, it must be remembered that not all infants with congenital heart disease have a heart murmur in the neonatal period.

- **A neonate with any of the following findings needs urgent assessment including echocardiogram even if a murmur is not present:** signs of heart failure or shock (see below), lower limb saturations <96% in the absence of respiratory disease, >3% difference between pre and post ductal saturations, absent/weak femoral pulses.

**The investigation of the neonate with a heart murmur (see attached flowchart)**

Investigation will vary depending upon local resources and expertise. The following recommendations represent the minimum requirements to ensure the safe management of neonates with heart murmurs and the timely identification of congenital heart disease.

- All infants with a heart murmur on neonatal examination should be reviewed by a senior paediatrician (middle grade or consultant).

- All infants with a heart murmur should remain in hospital until >24 hours old (unless definitive diagnosis is reached before this).

- All infant with a heart murmur should have a detailed cardiovascular clinical examination which must include measurement of pre and post ductal saturations.

- If a baby with a heart murmur is discharged before a definitive diagnosis is reached, the parents should be given a written information leaflet describing warning signs and advising them of what to do in the event that their baby became unwell.
**Clinical examination:**

- Dysmorphic features
- Signs of heart failure (tachypnoea, increased respiratory effort, hepatomegaly, shock)
- Palpation of brachial and femoral pulses
- Presence of cyanosis (as measured by lower limb saturations – a reading < 96% or >3% difference between pre and post ductal saturations should prompt further investigation 1)
- Heart sounds
- Presence of a heave
- Murmur – intensity, character, location and radiation

**Electrocardiogram**

- ECG has been shown to be a sensitive and specific tool for diagnosing atrioventricular septal defect2 (more common in infants with Trisomy 21) but has not been shown to aid significantly in the diagnosis of other structural congenital heart disease 3.

- A normal neonatal ECG shows right axis deviation because of the right ventricular dominance of the newborn heart. Left axis deviation in a newborn is a significant abnormal finding and should prompt further investigation.

- Whilst an abnormal ECG should prompt further investigation, a normal ECG should not be considered reassuring if there are abnormal clinical findings or lower limb saturations <96%.

**CXR and 4 limb blood pressure**

There is no evidence to support the use of CXR or 4 limb blood pressure measurements in the assessment of neonates with heart murmurs3,4,5,6.
**Echocardiography**

- This is the gold standard investigation for differentiating between innocent and pathological murmurs. Some units will undertake an echocardiogram in all neonates with heart murmurs. For many units this is not currently practical.

- In units where it is not feasible to perform echocardiogram for all infants with heart murmurs, information gathered from examination findings, oxygen saturations +/- ECG can be used to determine the need for and timing of echocardiography and follow up:

1. **Likely significant congenital heart disease – urgent echocardiogram and review (same day)**

   Infants with a heart murmur and any of the following warning signs: lower limb saturations < 96%; >3% pre / post ductal difference; absent/weak femoral pulses; signs of heart failure or shock. These infants require admission to a neonatal unit for consideration of prostaglandin and urgent discussion +/- transfer to a cardiac centre. If appropriately skilled local PEC or visiting cardiologist is available to perform echocardiogram while retrieval is awaited then this can be linked by telemedicine link / used to update surgical centre. **This should not be allowed to delay transfer.**

2. **Asymptomatic but clinically pathological murmur – soon echocardiogram (pre-discharge or as soon as possible within 1 week)**

   Infants without any of the above warning signs but with any of the following abnormal clinical findings: dysmorphism; heave; abnormal heart sounds; loud murmur (>2/6); pansystolic, diastolic, continuous murmur; murmur location other than left sternal edge /radiation; abnormal ECG.

3. **Low risk of congenital heart disease - routine review neonatal OPC 2-6 weeks**

   Well infants with no signs of heart failure, normal pulses, lower limb saturations >96%, soft (1-2/6) systolic murmur at the left sternal edge with no radiation.
References
(also see PECSIG “The investigation and management of neonatal heart murmurs: literature review.”

1. Impact of pulse oximetry screening on the detection of duct dependent congenital heart disease: a Swedish prospective screening study in 39 821 newborns. Anne de Wahl Granelli et al BMJ 2009;338;a3037


Based on the PECSIG 2013 guideline

Authors - PECSIG Neonatal Murmur Guideline Group
(Kathleen O’Reilly, Hilary Maddicks, Vishna Rasiah, Venu Gopalan with BCCA input from Rob Martin and John Simpson)

Adapted for local use by
Duncan Boyd, Maria Ilina, Brodie Knight and Kathleen O’Reilly.

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Appendices
1. Flow Chart
2. Parental Information Sheet
3. Full literature search
West of Scotland Neonatal Murmur Guideline

Heart murmur → suspect congenital heart disease

(Senior Paediatric Review
Cardiovascular Examination
Pre and post ductal saturations +/- ECG
Delay discharge until >24hours old)

FINDINGS
ANY of the following:
- Signs of heart failure/shock
- Lower limb saturations <96%
  >3% pre / post ductal difference
- Absent /weak femoral pulses

Significant congenital heart disease

LIKELY
ANY of the following:
- Loud murmur (>2/6)
- Heave
- Pansystolic/diastolic/continuous
- Location other than LSE
- Abnormal ECG (if done)
- Murmur + dysmorphic features

Congenital heart disease

URGENT
Admit to NICU - consider prostaglandin. Discuss with cardiac centre +/- Echo by PEC/ cardiologist whilst awaiting retrieval.

ACTION

CONGENITAL HEART DISEASE

SOON
Echo prior to discharge or review by cardiologist/ PEC as soon as possible within a week. Info sheet to all parents discharged prior to Echo.

Routine
Info sheet to parents prior to discharge. Paediatric review within 2-6 weeks +/- Echo if murmur still present.

Low risk of congenital heart disease

ALL of the following:
- Well baby
- No signs of heart failure
- Normal pulses
- Normal saturations
- Soft systolic murmur (≤2/6)
- Murmur heard at LSE only

Routine
Info sheet to all parents discharged prior to Echo.

Paediatric review within 2-6 weeks +/- Echo if murmur still present.
HEART MURMURS IN THE NEWBORN
INFORMATION FOR PARENTS

What is a heart murmur?

A heart murmur is an extra noise which is heard when the heart is listened to with a stethoscope.

Does a heart murmur mean there is heart problem?

No. Most babies with heart murmurs have completely normal hearts. These babies have what are known as “innocent” or “normal” heart murmurs. However, sometimes a heart murmur can be a sign that there is a problem with the heart like a small hole or a narrowing and this is why all babies with heart murmurs are reviewed.

How will I know if my baby has a heart problem?

Your baby will be seen in clinic within 2-6 weeks. If the murmur can still be heard and the doctor is not sure that it is an “innocent” or “normal” heart murmur then your baby will be referred to a heart specialist who may do further tests.

What should I look out for?

Most babies with heart murmurs remain well but if your baby becomes unwell they should be seen urgently by a doctor. Signs to look out for include: breathing difficulties; breathless or sweaty when feeding; poor feeding; blue colour of skin and lips or mottled skin.

What should I do if my baby becomes unwell?

You should seek urgent medical advice. Phone _____________________________. Explain that your baby has a heart murmur and has become unwell.

Points to remember

A heart murmur is an extra noise heard when listening with a stethoscope.
Most babies with heart murmurs have completely normal hearts.
A heart murmur can sometimes be a sign of an underlying heart problem.
IF YOUR BABY BECOMES UNWELL SEEK URGENT MEDICAL ADVICE.
The investigation and management of neonatal heart murmurs: literature review
Dr Kathleen O’Reilly, Dr Hilary Maddicks - March 2011

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Appendix – summary of individual papers

AIMS
The literature search was performed to enable the development of an evidence based guideline for the investigation and management of neonatal heart murmurs. In particular, the answers to the following questions were sought:

1. What are the causes and natural history of neonatal heart murmurs?
2. What evidence supports the use of the following investigations for neonatal heart murmurs: clinical examination; CXR; ECG; pulse oximetry; 4-limb blood pressure; echocardiography?

It is hoped that the guideline will:

1. Improve the pre-discharge diagnosis of life threatening cardiac malformations
2. Ensure early diagnosis and appropriate referral of significant cardiac malformations
3. Allow appropriate use of limited resources
4. Minimise repeat reviews and parental anxiety

METHODS
Pubmed literature search performed June 2010.

Limits activated: Humans, English, All infant: birth-23 months, All child: 0-18 years, Newborn: birth -1 month, publication date 01.06.1980 to 01.06.2010

Search terms:
Neonatal heart murmur investigation; Neonatal heart murmurs diagnosis; Neonatal heart murmur management; Neonatal heart murmurs clinical examination; Neonatal heart murmurs and echocardiogram; Neonatal heart murmurs and pulse oximetry; Pulse oximetry screening congenital heart disease; Measurement 4 limb blood pressure neonate; Neonatal heart murmur CXR; Neonatal heart murmur ECG.
RESULTS – SUMMARY OF EVIDENCE

Congenital heart disease and causes of neonatal heart murmurs


19.7% - 30% of infants with critical heart malformations are diagnosed post discharge of whom 75% are duct dependent systemic circulation (Mellander 2006, Wren 2008).

75% of babies who die of congenital heart disease are symptomatic within first 18 hours. 57% of infants dying of undiagnosed CHD post discharge had a murmur noted prior to discharge (Beebe 1996).

Neonatal heart murmurs are identified in 0.6-1.4 % of babies at neonatal examination (Ainsworth 1999, Farrer 2003).


Innocent heart murmurs are often associated with physiological branch PA stenosis which resolves in 2/3 of cases by 6 weeks (Arlettaz 1998).

Clinical examination


Experience neonatologists/ paediatricians are able to assess the significance of neonatal heart murmurs as well as paediatric cardiologists (Azhar 2006, Du 1997, Gokmen 2009).

Predictors of congenital heart disease include: harsh quality; location (RUSE, LLSE, apex) and timing (pansysolic, diastolic, continuous) (Mackie 2009).


With appropriate guidelines SHOs can accurately assess the significance of neonatal heart murmurs in 87% of cases (Farrer 2003).

Echocardiography


Echocardiography is not cost effective for screening of all babies for congenital heart disease (i.e those with no clinical signs of CHD) (Knowles 2005).

Early echocardiography may pick up PDA and PFO. An atrial septal defect <3mm in the neonatal period closes spontaneously in all cases before 9 months of age (Arlettaz 1998).

70% of babies undergoing echocardiography on the neonatal unit had either structural or functional cardiac abnormalities and initial echo findings directly influenced clinical management in > ¾ of babies (Moss 2003).

There is good concordance in echocardiographic findings between paediatric cardiologists and appropriately trained neonatologists (Moss 2003, Samson 2004).

**Pulse oximetry**


Has been proposed as a universal screening tool for congenital heart disease and is estimated to be cost efficient (Granelli 2009, Knowles 2005).

Published literature reviews the role of pulse oximetry in universal screening for congenital heart disease rather than specifically in subgroup of infants with neonatal heart murmurs although “the noting of a low saturation in addition to a murmur would almost certainly lead to an urgent rather than a routine referral” (Richmond 2002).

Median O2 value is 98.3% in 1st 24 hours (O’Brien 2000). Neonates need approximately 15 min to reach adult SpO2 values and functional occlusion of the ductus arteriosus occurs in the 1st 20 mins of life (Toth 2002).

Pre and post ductal pulse oximetry can detect 100% of infants with pulmonary duct dependent circulation and 50 - 85% of left heart obstructive lesions (Granelli 2009, Hoke 2002).

Clinical examination combined with pulse oximetry can detect 92% of infants with duct dependent congenital heart disease (Granelli 2009).

A single lower extremity reading with cut offs 95-96% is the most appropriate for large-scale screening (Mahle 2009, Valmari 2007).

Addition of upper and lower measurements would increase the sensitivity of pulse oximetry without a decrease in specificity (Granelli 2005 quoted in Mahle 2009, Thangaratinam 2007).

**Chest X-Ray**


Most of published literature refers to children with heart murmurs rather than neonates.

Swenson 1997 found that 67% of patients with definite cardiac pathology have a normal CXR and ECG. Although Swenson concluded that CXR was useful in children with a murmur or chest pain the diagnosis of “no heart disease” was not confirmed by echocardiography.

CXR is of no help in the discrimination between heart disease and no heart disease in asymptomatic children examined by a paediatrician or referred to a cardiologist for a cardiac murmur (Birkebaek 1995, Macleod 2001, Newburger 1983)
Although experience radiologists achieve high accuracy in differentiating cardiac form non-cardiac disease, neonatal chest radiographs are unlikely to provide the definitive diagnosis and cannot be recommended in the initial evaluation of the asymptomatic neonate with a heart murmur (Oeppen 2002).

**ECG**


ECG is a sensitive and specific test for diagnosing cAVSD in infants with Trisomy 21 (Narchi 1999) but may still miss 17% of cases (Tubman 1991).

18% of children with ASD have a normal ECG (Christensen 2005).

67% of children with definite cardiac pathology have a normal CXR and ECG (Swenson 1997) yet paediatricians may consider a normal ECG as confirmation of a clinical diagnosis of an innocent murmur (Macleod 2001).

Although Swenson concluded that ECG was useful in children with a murmur or chest pain the diagnosis of “no heart disease” was not confirmed by echocardiography.

**4 limb blood pressure**

Relevant papers: Crossland 2004, Rahiala 2001

There is very little published evidence about 4 limb blood pressure measurements in neonates. Although cardiology textbooks advocate its use when coarctation is suspected they fail to define what would constitute an abnormal result (Pediatric Cardiology:The Requisites in Pediatrics, Victoria Vetter, Mosby Elselvier 2006).

In children/ adults a cut off point to warrant further examination = 5-10mmHg higher in the arm than the leg (Rahiala 2001).

4 limb blood pressure does not help confirm or exclude coarctation of the aorta and has an 8% false positive rate which would result in 300 urgent cardiac opinions for each correct diagnosis made (Crossland 2004).
CONCLUSIONS/RECOMMENDATIONS

1. All neonates noted to have a heart murmur should be examined by an experienced paediatrician.
2. All neonates with a heart murmur should have pre and post ductal saturations measured.
3. All neonates noted to have a heart murmur should remain in hospital until 24 hours old unless a definitive echocardiographic diagnosis is reached before this time.
4. There is no evidence to support the routine use of CXR, ECG or 4 limb bp in the assessment of the asymptomatic neonate with a heart murmur.
5. In cases where an ECG has been performed, a normal result should not be considered confirmation of an innocent murmur.
6. Echocardiography remains the gold standard investigation for neonatal heart murmurs.
7. Clinical examination findings and pulse oximetry should be used to inform decisions about the timing of review +/- need for urgent echocardiography.
8. Parents of neonates with heart murmurs who are discharged prior to definitive diagnosis should be given written information about warning signs and advised who to contact if concerns.
SUMMARY OF INDIVIDUAL PAPERS

The following papers were reviewed:


All newborn infants undergoing neonatal examination < 48 hours. 0.6% babies found to have a murmur @ routine examination of whom 54% had a structural cardiac malformation.

9% of babies with murmurs required early cardiac surgery.

"Early referral of all symptomatic babies with murmurs is recommended"


Healthy term newborn infants with a murmur (thought to be innocent at initial examination). 50 babies/ 50 controls. Echo and follow up at 6wks, 3 months and 6 months.

Innocent murmurs prevalence 21/1000. 42% of babies with innocent murmur <1 day old.

All babies had PFO at 1st assessment. 66% still had PFO at 6 weeks of whom 52% had murmur. At 3 months 3 babies still had PFO. An atrial septal defect < 3mm in neonatal period closes spontaneously in all cases before 9 months of age.

PDA seen in 80- 90% of babies < 24 hours old but had closed in all cases by 6 weeks.

Innocent heart murmurs persisting > 24 hours age often related to physiological pulmonary branch stenosis which resolves in 2/3 cases by 6 weeks and in all cases by 6 months.


Saturation cut off $\geq 95\%$.

Sensitivity/specificity of pulse oximetry for cyanotic heart disease – 100%/ 99.7%.

Recommend referral if sats $< 95\%$.


75 neonates < 28 days referred to cardiologists by neonatologists.

Based on clinical examination murmur classed as innocent/ possibly pathological/ pathological by neonatologist and cardiologist. Diagnosis confirmed by echo.

Clinical examination by neonatologist: Sensitivity 78%; specificity 33%; PPV 77%; NPV 37%.

Clinical examination by cardiologist: sensitivity 83%; specificity 25%; PPV 80%; NPV 29%.

Most common pathological murmurs PDA, VSD, ASD,TGA.

"Certified neonatologists are able to assess the significance of neonatal heart murmurs as well as paediatric cardiologists."

"Echo still needed to reach the accurate diagnosis of CHD in neonates even if a paediatric cardiologist is consulted."

**Bakr A, Habib H.** Combining Pulse Oximetry and Clinical examination in Screening for Congenital Heart Disease. *Pediatr Cardiol* 26:832-835, 2005
5211 infants mean age 31.7 hours. Saudi Arabia.
Pulse oximetry right upper and lower limbs. Fractional oxygen saturation ≥ 94% considered normal. < 90% - echo. 90-94% - verified by 3 readings - echo. All murmurs ≥ 2/6 referred for echo.
15 echos performed: 5 due to saturation screening (TAPVR, PA, VSD, TA, normal); 10 owing to murmurs (2 PDA, 3 VSD, 1 ASD, 4 normal)
3 cases not picked up by screening: CCTGA, PS, VSD.
Pulse oximetry alone: sensitivity 31%, specificity 100%, PPV 80%.
Examination alone: sensitivity 46%, specificity 100%, PPV 60%.
Pulse oximetry and examination combined: sensitivity 77%, specificity 100%, PPV 66.7%.
"Relatively low sensitivity may limit the ability of screening method to identify all cases of CHD. However, high specificity would diminish parent anxiety and obviate unnecessary echo and costly follow up.”

4/7 (57%) of infants who died from CHD post discharge had murmurs noted prior to discharge. 75% of infants who died from CHD were symptomatic before 18 hours of age.

Birkebaek NH, Hansen LK, Oxhoj H. Diagnostic value of chest radiography and electrocardiography in the evaluation of asymptomatic children with a cardiac murmur. Acta Paediatr. 1995 Dec;84(12):1379-81 Abstract only
100 children with a murmur (1 month -15 years median age 30.1 months) Physical examination + ECG + CXR.
After physical examination patients divided into: no heart disease/ probable heart disease/ heart disease. Diagnosis re-evaluated following CXR and ECG. Diagnosis incorrectly changed in 3 children following ECG and CXR.
"CXR and ECG is of no help in the discrimination between heart disease and no heart disease in asymptomatic children referred for a cardiac murmur.”

120 children referred to paediatric cardiologist with murmurs. (4 days – 14 years median age 10 months)
Divided into 3 groups based on clinical examination / ECG: no heart disease; possible heart disease; definite heart disease.
Sensitivity/ specificity of physical examination 90.3%/93.8%.
3 cases diagnosis incorrectly changed on basis of ECG.
"ECG is of no help in the discrimination between heart disease and no heart disease in children referred to the cardiologist for a murmur.”

Retrospective review of 47 patients undergoing ASD closure (1 month – 17 years)
18% had normal ECG.
78% had right ventricular conduction delay.
58% had right axis deviation.
13% had right atrial enlargement.
No statistical correlation between number of physical and ECG findings and size of defect, age at presentation or body surface area.


40 infants enrolled (38 measurements performed) age 2-21 days. Normal arch and closed duct on echo. Positive test defined as > 20 mmHg between nearest arm bp and nearest leg bp. Wide variation in bp between limbs when single readings are taken using oscillometric or Doppler devices. False positive rate 8% = 300 urgent cardiac opinions for each correct diagnosis made. “Clinical findings should raise suspicion of CoA sufficiently to prompt referral and 4 limb bp does not help to confirm or exclude CoA.”


116 neonates with heart murmurs persisting > 24 hours (12 hours – 14 days; 90 term, 26 preterm). 97/116 (84%) had CHD (includes PDA)
Following clinical examination babies categorized as: definite heart disease; possible heart disease; innocent murmur. Diagnosis confirmed by echo.
88/116 neonates clinically felt to have definite heart disease: 10% changed to normal after echo. 10% lesion specific diagnosis changed.
5/21 (24%) neonates with possible congenital heart disease: diagnosis changed to normal heart after echo.
1/6 neonates with “innocent murmur” changed to VSD following echo.
90% of heart diseases diagnosed clinically were confirmed by echo. 96/97 neonates with heart disease were referred by paediatrician as definite or possible heart disease.
“Clinical examination by an experienced paediatrician is the most important means of assessing neonates with heart murmurs. Lesion specific diagnosis is not satisfactory. Echocardiography is necessary for neonates with clinically diagnosed congenital heart disease.”


Population: all babies on PNW who had a routine neonatal examination by an SHO. Guideline split babies with murmurs into 4 groups:
Cyanosis, heart failure, poor pulses Admit NICU
Positive family history/ concerning antenatal scan discuss with senior colleague
≥ 3/6, harsh, diastolic, pansystolic, continuous, click next cardiology clinic
Clinically innocent murmur non-urgent referral to neonatal cardiac clinic
112 (1.4%) of babies found to have murmur on routine neonatal examination. 90 babies had an echo of whom 23% had an abnormality. Mean waiting time for clinic 9 weeks (7 weeks for clinically significant murmurs/10 weeks for clinically innocent murmurs)
Murmur assessment by SHO: sensitivity 71%, specificity 91%. SHOs accurately assessed the significance of the murmur in 87% of babies.
“Given appropriate guidelines, SHOs can assess the clinical significance of neonatal murmurs and decide upon appropriate follow up.”

58 infants with a heart murmur and 59 controls. Turkey. Divided into innocent/pathological based on clinical examination and diagnosis confirmed by echo.

CHD identified in 19/53 (35.8%) of infants with a murmur. Most common cause of pathological murmurs: PDA, ASD, VSD, PS.

Examination by paediatrician: 33.3% sensitive, 95.5% specific.

Examination by cardiologist: 40% sensitive, 98.9% specific.

“A trained paediatrician can assess the significance of and determine appropriate management for neonatal heart murmurs as well as a paediatric cardiologist. Echocardiography is still required for accurate diagnosis of CHD in neonates even if a paediatric cardiologist is consulted.”


Screening cut-off values: < 95% saturation or > 3% pre/ post ductal difference.

Recommend pre and post ductal screening as post ductal only may miss complex lesions e.g. transposition with arch obstruction.

Introduction of pulse oximetry should be at a minimum cost neutral.

Pulse oximetry (pre and post ductal) detects:

- 100% of infants with pulmonary duct dependent circulation
- 50% of left heart obstructive lesions
- 92% of all infants with duct dependent circulation when used in combination with clinical examination.

Green K, Oddie S. The value of the postnatal examination in improving child health. *Arch Dis Child Fetal Neonatal Ed* 2008; 93: F389-F393

Minimising repeated reviews before a final diagnosis is reached or excluded reduces parental anxiety (from hearing screening).

Good quality initial explanations may reduce the anxiety implicit in any repeated review. (Giuffre Canadian Journal of Cardiology 2002)


587 neonates referred to rapid access murmur clinic over 4 year period.

Seen by paediatric cardiologist at mean age of 25 days (1-90)

None had life- threatening cardiovascular malformation.

411/587 (70%) normal; 11/587 (2%) had significant cardiovascular malformation; 127/587 (22%) had minor cardiovascular malformation.

In same birth cohort, 248 babies with CVMs were referred through other channels: 64 life threatening; 78 significant and 106 minor CVM.

Rapid access murmur clinic led to a diagnosis of: 0% of life-threatening CVM; 12% of significant CVM; 56% of minor CVM.

“The rapid access murmur clinic does not contribute to the identification of life-threatening CVM.”

2876 well newborns screened and 32 infants with known congenital heart disease
Saturation screening: abnormal < 92% in air or 7% lower in leg than arm.
2876 well newborns in nursery: 57 (0.02%) had abnormal saturation screening or whom 4/57 had congenital heart disease.
Of the 32 babies with congenital heart disease 11/13 (85%) of those with left heart obstruction had abnormal results and 15/19 (79%) of those with other forms of congenital heart disease.


Additional cost per timely diagnosis of life-threatening CHD: £4900 for pulse oximetry; £4.5 million for screening echocardiography.
Additional cost per additional timely diagnosis of significant CHD: £1500 for pulse oximetry; £36,000 for screening echocardiography.
Screening echocardiography is associated with the highest detection rate but is the most costly strategy and has a 5% false positive rate.


Effectiveness of pulse oximetry screening for critical CHD (would require surgical correction within 1st month) in asymptomatic newborns.
11,281 asymptomatic newborns (excluding prenatally diagnosed). Abnormal result defined as postductal sats \( \leq 95\% \).
3 cases of critical CHD detected: 2 TAPVD, 1 TA. (1/3760 asymptomatic newborns found to have critical CHD by oximetric screening)
1 false positive – PPHN
2 cases missed: 1 CoA, 1 hypoplastic left PA with AP collaterals.
Sensitivity 60%, specificity 99.9%, PPV 75%, NPV 99.8%.
"Oximetric screeing for critical CVM compares favourably to other newborn screening programs already in place”


201 neonates with murmurs (> 24 hours \( \leq \) 31 days median age 12 days) examined by paediatric cardiologist.
Assessed diagnostic accuracy of clinical examination/ECG in differentiating between pathological and non-pathological murmurs.
56% of neonates with murmurs had congenital heart disease (not clear if all murmurs sent to this clinic or only those that paediatricians thought were pathological).
Clinical examination alone: 80.5% sensitivity; 90.9 % specificity.
Following factors independent predictors of CHD: harsh quality, location, timing.
ECG did not improve the sensitivity of the clinical assessment in the detection of CHD (note no infants with AVSD in study group).

Macleod C. Evaluating cardiac murmurs; are diagnostic tests helpful? Irish Medical Journal 2001 May; 94(5):154-5

81 children > 1 year with murmurs examined by paediatricians
The routine use of CXR and ECG in the evaluation of children > 1yr with murmurs does not help general paediatricians to distinguish between innocent and pathological murmurs. “It appears that normal ECG and CXR findings are taken as confirmation of a clinical diagnosis of innocent murmur.”


Median value of sats in healthy term infants is 97.8% at 20-24 hours of age and 97.6% at 2-7 days (Poets 1999).
Using cut off of 95% in lower limbs 81% of infants with CCHD could be identified. (Hoke 2002).
Oximetry screening before 24 hours of life can result in 5% false positive rate. If done at time of discharge false positive rate falls to 1%. (Thangaratinam et al 2007)
The mean difference between SpO2 in upper and lower extremities is < 1%.
Differences in SpO2 of more than 3% or 4% might be used to identify newborns with CCHD who might otherwise be missed. (Reich 2003, Granelli 2005)
Addition of upper and lower measurements would increase sensitivity from 89.4% to 92.4% without a decrease in sensitivity. (Granelli 2005. Study done in infants with known CCHD)
10 studies examining oximetry screening for CCHD: 123,846 infants. 3 studies ≥ 96%, 5 ≥ 95%, 1 ≥ 94%, 1 ≥ 92. 3/10 studies performed pre and post ductal sats (cut off ≥ 4/7 difference). 0.87 false positive rate (0.035% false positive rate if > 24 hours old).
“A single lower-extremity reading would appear to be the most appropriate for the purposes of large-scale screening.”
“When neonates are identified as having hypoxaemia (SpO2 ≤ 95% it is necessary to evaluate them for CCHD.”


Retrospective review of all children between 1993-2001 who had surgery/catheter intervention before the age of 2 months.
0.09% of all infants had critical CHD (50% duct dependent systemic circulation, 40% duct dependent pulmonary circulation, 9% not duct dependent)
Duct dependent systemic circulation diagnosed later (CoA later than HLHS, AS, IAA)
19.7% diagnosed post discharge of whom 43% were in shock at time of admission and 75% were duct dependent systemic circulation.


Infants undergoing echocardiography on the neonatal unit – 157 echos in 82 infants.
Most common reason for performing an echo was asymptomatic murmur.
54% of babies had a definite structural cardiac abnormality identified on echocardiography.
70% of babies had either a structural or functional cardiac abnormalities.
Initial echo findings directly influenced clinical management in over ¾ of babies.
In 31/38 infants who had paired scans by neonatologist and cardiologist there was complete concordance. In 7/38 there were minor discrepancies which did not prevent appropriate clinical management.
“Study supports the widespread current practice in the UK of neonatologists performing echocardiograms”.

Prospective study of value of routine neonatal ECG to diagnose CHD in Down Syndrome. 37 newborns with Trisomy 21 Jan 87 – Dec 96.
13/37 (35.2%) had CHD. 5/13 (38.4 %) had AVSD
Left axis deviation to diagnose cAVSD in Down syndrome: 100% sensitivity, 96.8% specificity, PPV 83.3%, NPV 100%.
Superior axis in Down syndrome: 100% sensitivity, specificity, PPV, NPV for AVSD.
The abnormal axis, when present at birth, did not change subsequently, and, if absent at birth, did not appear later (abnormal QRS plane is related to congenital alterations of the excitation pathways into the ventricles).
“The predictive value of a neonatal ECG to diagnose heart defects other than AVSD in asymptomatic neonates with Down syndrome is therefore very poor”.


280 children > 1 month with heart murmur examined by a cardiologist.
“The results of diagnostic tests are unlikely to change the clinical diagnosis of no heart disease or definite heart disease, when made by a qualified paediatric cardiologist in children newly referred for evaluation of a heart murmur.”

**Norman M.** Detecting heart defects in newborn infants – innocent murmurs mixed with silent dangers. *Acta Paediatrica* 2006; 95:391-393


90 infants > 37 weeks gestation < 24 hours old. Continuous sats monitoring (? where sats probe located. Nellcor ? functional or fractional)
Median O2 value is 97.8% @20-24 hours
Median O2 value is 98.3% in 1st 24 hours ( range 89-100)
0-4 hours 99%
4-8 hours 98.4%
8-12 hours 98.6%
12-16 hours 98.1%
16-20 hours 98.3%


68 asymptomatic neonates with murmurs.
Retrospective review of CXR.
Chest radiograph did not influence clinical management. Although radiologists who frequently report neonatal chest radiographs achieve high accuracy in differentiating cardiac from non cardiac disease, chest radiograph unlikely to provide the definitive diagnosis.
“Chest radiographs cannot be recommended in the initial evaluation of the asymptomatic neonate with a heart murmur”.
**Patton C, Hey E.** How effectively can clinical examination pick up congenital heart disease at birth? *Arch Dis Child Fetal Neonatal Ed 2006; 91:* F263-F267

14, 572 babies undergoing routine clinical examination between 1996 and 2003. If a murmur present then sats performed and review arranged within 1st week. Cardiac assessment before discharge if breathless, poor pulses, signs of LVOT obstruction or sats <90%.
176/14,572 (1.21%) found to have a structural heart defect before age of 1 year. 150/176 suspected before discharge.
In 2000-2003 80% diagnosed before 6 weeks of age.
Parents did not seem to find the delay in the screening system unduly stressful as long as they were properly informed of what is going on and why. (in contrast to finding by Giuffre 2002 that “the time parents spend waiting for a cardiac diagnosis often generates more stress than the diagnosis itself”).

**Rahiala E, Tikanoja T.** Suspicion of aortic coarctation in an outpatient clinic: How should blood pressure measurements be performed? *Clinical Physiology 21, 1,* 100-104

Population: 40 healthy children (11.0 +/- 0.4 years), 20 healthy students (18.8 +/- 0.9 years), 19 children with previous coarctation repair (9.3 +/- 4.0 years)
Measurement of bp in arms, thighs and ankles using sphygmomanometer oscillometric device. Successive measurements in the same limb resulted in lower systolic values than the 1st measurement.
In patients with coarctation gradients obtained correlated well with echo gradients.
Healthy children: arm bp = thigh bp -2mmHg= ankle bp -12 mmHg
Adults: arm bp = thigh bp - 12mmHg = ankle bp -16mmHg
Cut off point to warrant further examination = 5-10mmHg higher in arm than leg.
Ignore 1st bp then use repetitive measurement in the arm with the higher systolic bp.
The possibility that the right brachial artery leaves the aortic arch after the left brachial artery and distal to a coarctation should be ruled out with measurements in both arms.


Single pulse oximeter reading before discharge (normal >94%)
2114 infants enrolled: 88 (3.8%) echos performed, 43/88 abnormal 12 required management. 2851 births in control group: 108 echos, 42 abnormal, 13 required management.
“Routine pulse oximetry was nearly 100% specific for detecting cyanotic CHD. Routine pulse oximetry did not lead to an increase in the number of echocardiograms.”

**Reich J, Connolly B, Bradley G et al.** The reliability of a Single Pulse Oximetry Reading as a Screening Test for Congenital Heart Disease in Otherwise Asymptomatic Newborn Infants. *Pediatr Cardiol (2008) 29:885-889*

7962 infants born of whom 12 had duct dependent heart disease. None were initially identified by saturation screening (normal >94%) although 4/12 failed. 1450 pulse oximetry traces retrospectively reviewed - < ½ can be demonstrated to be reliable.
“A nurse with a degree of LPN or higher placing the probe for 360 seconds can optimize reliability at >95%.”
Conclusion: before pulse oximetry utilised as a screening test, adequate training of staff necessary and need a large research study to verify that the sensitivity of the test justifies its use for universal screening.

Israel 1994-1996
Term newborns < 5 days with an asymptomatic heart murmur. Echo performed within 12 hours (median 4 hours)
170 newborns with murmurs: 86% had CHD including PDA; 68% excluding PDA.
2/3 had a L R shunt, 37% had VSD, 23% had PDA
23/170 had a diagnosis with a significant clinical implication.
5/170 had a life threatening abnormality.
Chance of finding a cardiac defect in asymptomatic babies with a murmur is significant.
L R shunt and VSD may cause a murmur even on the 1st day of life.
Any newborn with a murmur should undergo echo in the 1st few days of life.

Richmond S, Reay G, Abu Harb M. Routine pulse oximetry in the asymptomatic newborn. *Arch Dis Child Fetal Neonatal Ed* 2002;87: F83-F88

Population: 6166 inborn infants.
Fractional O2 sats - cut off <95%
5% of babies had sats < 95%. 1 %of babies had 2 sats readings <95%.
Sensitivity for detecting CHD (excluding VSD) 53% and specificity 99%.
Measuring sats was possible in 98% of eligible infants.
"If an echocardiogram is not immediately available, then the noting of a low saturation in addition to a murmur would almost certainly lead to an urgent rather than routine referral."

Rivera I, Mecdonca da Silva M, Fernandes J. Congenital Heart Diseases in the Newborn: from the Pediatrician’s Request to the Cardiologist’s Evaluation. *Arq Bras Cardiol* 2007; 89(1): 6-10

358 newborns referred for cardiac evaluation (9.6% of births).
400 random newborns also evaluated.
181/358 normal
128/358 (36%) PDA
49/358 CHD
256/358 (72%) referred because of a murmur
39/256 (15%) of murmurs had CHD
117/256 had PDA
90/256 (35%) normal
10/256 (4%) PFO

Samson G, Kumar S. A study of congenital cardiac disease in a neonatal population – the validity of echocardiography undertaken by a neonatologist. *Cardiol Young* 2004;14: 585-593

11,085 liveborn infants
Incidence of congenital cardiac malformations 7.49/1000.
Babies with murmur persistent at 48 hours referred for echo.
165 infants referred for echo (68 with asymptomatic murmur, 11 with symptomatic murmur, others included arrhythmia and infant of diabetic mother)
59/68 had echo (median age 4 days range 2-10)
19/59 murmur had gone by time of scan – all had normal scans
40/59 murmur still present at time of scan and 30/40 had a defect
Concordance between the neonatologist and cardiologist was good – Cohen’s Kappa coefficient 0.68 95% confidence interval 0.51-0.85.
"Safeguards need to be put in place - training and accreditation and appropriate guidelines for referral. Ultimate management of an infant with a cardiovascular malformation is the domain of the paediatric cardiologist."

161 children 1 month - 17 years (median 3.2 years)
Examination by paediatric cardiologist + ECG. Patients divided into 3 groups: pathologic/possibly pathologic/innocent
Clinical examination by a paediatric cardiologist: 96% sensitive, 95% specific, PPV 88%, NPV 98%.
No diagnosis changed after electrocardiography.
"Clinical examination by a paediatric cardiologist is the most useful means of initial evaluation of referred pediatric murmurs."
"ECG may assist in reaching a definitive diagnosis only in those patients suspected to have heart disease."


106 children 1 month – 14 years
79% of patients referred with heart murmur/ 21% with chest pain.
Examination by cardiologist led to initial diagnosis of no heart disease 60/106 possible heart disease 25/106 and definite heart disease 21/106.
4/60 diagnosis changed from no heart disease to definite heart disease – 3 patients with abnormal ECG ( ASD – RVH on ECG, HCM – LVH on ECG, WPW) and 1 patient with abnormal CXR – partial absence of pericardium.
NB .53/60 in the no heart disease did not have echo confirmation of that diagnosis.
5/25 patients with possible heart disease changed to definite following ECG and CXR and 7/25 changed to no heart disease ( 6/7 had an echo!).
"ECG and CXR helped to diagnose heart disease in 4 patients thought to have no heart disease, helped to rule out lesions in 7 patients with possible heart disease (???? How), helped diagnose heart disease in 5 patients thought to have possible heart disease and helped confirm heart disease in 9 patients"
67% of patients with definite cardiac pathology do not have abnormalities in either CXR or ECG.


Sensitivity 25-98.5%
High specificity 98-100%
False +ve 0-2%
Highest sensitivity: threshold level < 95% or 3% difference between foot and right hand
Sensitivity higher in studies screening for critical or cyanotic heart disease.
Highest specificity when baby > 24 hours old.


50 neonates – 46@ 38-43 weeks, 3@ 37 weeks, 1@ 35 weeks

<table>
<thead>
<tr>
<th>SpO2</th>
<th>2 min</th>
<th>5 min</th>
<th>10 min</th>
<th>15 min</th>
<th>20min</th>
</tr>
</thead>
<tbody>
<tr>
<td>preductal</td>
<td>73(44-95)</td>
<td>84 (48-99)</td>
<td>92(65-99)</td>
<td>94(77-100)</td>
<td>95(78-100)</td>
</tr>
<tr>
<td>postductal</td>
<td>67(34-93)</td>
<td>78(42-97)</td>
<td>89(62-99)</td>
<td>92(71-100)</td>
<td>94(95-100)</td>
</tr>
</tbody>
</table>
Mean preductal SpO2 at 2 and 5 mins significantly higher than post ductal p< 0.05. By 10 mins not statistically significant.
Threshold of 95% SpO2 was achieved after 12 min preductal and 14 mins post ductal.
Conclusions: Neonates need approx 15 min to reach adult SpO2 values. Functional occlusion of the ductus arteriosus occurs in the 1st 20 in of life.

81 newborn infants with Down syndrome. 34 had CHD of whom 13 had AVSD.
17% of AVSD missed by ECG.
39% of AVSD missed by CXR.
15% of AVSD missed by ECG, CXR and examination together.
10/13 babies with an abnormal ECG had AVSD (1 VSD, 1 ASD and 1 large PDA)
ECGs may be abnormal particularly in the presence of an AVSD. An abnormal electrocardiogram had a high positive predictive value for congenital heart disease (1.0).

**Valmari P.** Should pulse oximetry be used to screen for congenital heart disease? Arch Dis Child Fetal Neonatal Ed 2007; **92:** F219-F224
Review of 10 studies (44, 969) evaluating pulse oximetry screening.
Pre/post ductal sats 73%/67% at 2 mins and 92%/89% at 10 mins – Toth 2002
In every newborn both measurements were 95% within 1 hour – Toth 2002
The use of SpO2 for screening is not practical before the age of 1-2 hours. Thereafter SpO2 is stable at around 98%.
Byrne 1995: Cyanotic heart disease - SpO2 <88% or pre/post ductal difference of ≥ 7%. Duct dependent left heart lesions- pre and post ductal difference of 4-5%
Sensitivity of pulse oximetry screening in serious CHD was 72%.
Cut offs of 95-96% seem high enough to make the measurement of a preductal/postductal SpO2 difference unnecessary.

Cardiovascular malformations 6.4/1000 of which 15 % were life threatening.
Of the life threatening malformations: 8% diagnosed prenatally; 62% diagnosed postnatally predischarge; 25% diagnosed postnatally post discharge; 5% diagnosed at post-mortem (none in previous 6 years).
Malformations most likely to remain undiagnosed at discharge: coarctation 54%; IAA 44%; AS 40%; TAPVD 37%.
Discussion:
It is impractical to refer all neonates with murmurs for urgent paediatric cardiology assessment.
If a significant murmur is detected and echocardiography skills are available in the neonatal unit, earlier diagnosis of potentially serious cardiovascular malformations ought to lead to a better outcome.