What is the role of fetal echocardiography (2D 76825, cardiovascular color flow mapping 93325) as performed in conjunction with detailed fetal anatomy scan (CPT 76811) now that AIUM requires limited outflow tract views?

I. Fetal echocardiography as performed and interpreted by MDs subspecialty trained and accredited in the recognition and diagnosis of fetal cardiac disease remains the evidence-based “gold standard” to achieve 90-100% detection of congenital heart disease (CHD) and to complete and optimize risk assessment for aneuploidy and nonchromosomal syndromes.

It is not the mere performance of “outflow tract” imaging but the detailed knowledge of fetal cardiac pathology, its recognition, and the interpretative expertise that is necessary to substantially increase detection rate of CHD.


Summary –

CPT 76805 – Nondetailed Obstetrical Ultrasound- four chamber and proximal outflow tract views
  _ 40-50% CHD detection.
  _ no evidence based Down syndrome (DS) detection

CPT 76811 – Detailed ‘level II’ Obstetrical Ultrasound- four chamber, proximal and mid outflow tracts views.
  _ less than 60-70% CHD detection.
  _ 50% DS detection.

HVRA’s Cardio Vascular Genetic Detailed L2 Obstetrical Ultrasound.
CPT 76811, 76825, 93976
  _ 90% CHD detection.
  _ 90% DS detection.
Outcome analysis of HVRA’s ‘universal’ fetal cardiac ultrasound program – 76811 with 2D fetal cardiac and cardiovascular color flow mapping 76825, 93325

- 10-year retrospective analysis of transfer rates of sick newborns with diagnosis codes of congenital heart disease delivered at Nyack Hospital demonstrated a transfer rate of 3-4 per 3,000 deliveries – significantly lower than the national average of 9-15 per 3,000 for similar-sized hospital (statistics obtained from Center for Disease Control.)

- 2008 prospective detection rate of cardiovascular disease
  - 22 malformations were identified in the 2,718 cases studied for an incidence of 2.4 cases per 300 newborns.
  - This compares to the most frequently quoted incidence of clinically significant congenital heart disease amongst newborns of 1:200 – 1:300.

II. AIUM’s updated limited cardiac outflow tract protocols – without fetal echocardiography – in the detection of fetal congenital heart disease, a literature review.

Interpretation of published studies must take into account the following factors that influence sensitivity (detection rate) of congenital heart disease.

- Population studied – unselected versus low risk.
  - Unselected population includes high-risk pregnancies including aneuploidy and/or extracardiac malformations.
  - Low-risk population – attempts to exclude patients with traditional pretest risk factors i.e. family history, maternal disease, drug disclosure.

- Gestational age at time of diagnosis – studies including CHD detection after 24 weeks exaggerate clinical value.

- Single center versus multicenter studies – multicenter studies better reflect cross-sectional clinical practice.

There are no published prospective multicenter studies that establish the sensitivity of screening four-chamber and outflow tract protocols in the population in which congenital heart disease is most prevalent – in low-risk euploid pregnancies with no extracardiac malformations – the latter of which would increase cardiac scrutiny i.e. falsely increase screening sensitivity.
Background – the American published data prior to AIUM institution of outflow tract protocol.


A retrospective 10-year statewide (Utah) surveillance study of 1,474 cases of congenital heart disease between 1997-2007.
  - Majority of CHD cases were missed prenatally and detection rates did not increase substantially during study period.
  - 25% of CHD was missed amongst mothers who had one or more MFM ultrasound studies.

European literature review of CHD detection with four-chamber and outflow tract protocols but not fetal echocardiography.


From Italy, in a multicentered prospective study of 4,873 low-risk patients –
  - 64% sensitivity for CHD (39 cases).
  - Operators received specialized training.
  - Gestational age at time of detection not stated.


From Norway, in a prospective single center of greater than 30,000 unselected patients examined at 18-20 weeks –
  - 44% sensitivity for isolated CHD
  - 67% sensitivity for CHD in fetuses with abnormal karyotype and/or extracardiac malformations.
Most recent American publications on congenital heart disease detection with four chamber and outflow tract views – but not fetal echocardiography.


○74% detection rate amongst 93 patients with significant congenital heart disease from a low-risk population yielding an incidence of 3.6 per 1,000 live births.

  ▪ The live birth incidence of significant CHD quoted in this study is less than the 6 per 1,000 moderate-to-severe congenital heart disease cases expected based on an analysis of 62 studies published after 1955. J Am Coll Cardiol 2002 June 19; 39(12): 189-1900.

○This article suggests the possibility of improvement in CHD detection employing outflow tract in comparison to the well-documented 30-50% detection rate in developed countries.

  ▪ This study was performed within an idealized environment requiring dedicated training and multidisciplinary review of screening studies.

  ▪ The 74% detection rate included fetuses with other extracardiac malformations and aneuploidy necessitating increased scrutiny of the heart.

○Therefore even under idealized conditions a lower CHD detection rate could be expected for the majority of fetuses with CHD, i.e. as an isolated lesion in patients with no risk factors.
III. **AIUM outflow tract protocol as a screening tool is not intended to nor does it achieve the clinically most important interpretive diagnostic endpoint** – reporting the exclusion of the five major patterns of congenital heart disease allowing safe delivery in a community hospital – hypoplastic left heart, Tetralogy of Fallot, transposition of the great vessels, atrioventricular septal defect, aortic coarctation.

- AIUM’s new limited outflow tract protocol remains an incomplete cardiovascular evaluation which does not include the following:
  - Transverse views of the aortic arch for laterality and coarctation
  - Evaluation of the cardiac valves for regurgitation and stenosis
  - Superior/inferior vena cava and pulmonic veins for anomalous pulmonary venous return
  - Right subclavian artery – see below.

IV. **AIUM’s updated 76811 limited outflow tract protocol remains an incomplete exam for genetic and syndromic evaluation.** Detailed obstetrical ultrasound without 2D cardiac 76825 and cardiovascular color flow mapping for genetic views has a 50-70% Down syndrome detection rate in comparison to a 90% detection rate when echocardiography is included.

The following vascular pathologies as aneuploidy and/or syndromic markers – are missed on AIUM’s detailed 76811 protocol.

- Identification of an aberrant right subclavian artery and identification of normal course to the right subclavian artery has positive and negative Down syndrome likelihood ratios of 27 and 0.7 respectively
- Agenesis of ductus venosus
- Aberrant drainage patterns of ductus venosus and umbilical vein
- Mitral and tricuspid valve regurgitation

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