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| **Prenatal Detection of Severe Structural Congenital Heart Defects** | |
| This metric will serve as a means for centers to track and report rates of prenatal detection of severe structural congenital heart defects, defined as lesions requiring surgical or catheter intervention within first 28 days of life. | |
| Numerator | Number of patients who had a prenatal diagnosis of structural congenital heart defect (CHD) in which intervention was expected or possible |
| **Denominator** | All patients undergoing initial surgical or catheter intervention for a structural congenital heart defect at ≤ 28 days of life |
| **Period of Assessment** | Quarterly |
| **Sources of Data** | Means of data collection will be center specific but may include:  Medical record/Chart review  STS or other surgical registries or databases  IMPACT or other catheterization registries or databases |
| **Rationale** | |
| Congenital heart defects account for the largest percentage of birth defects that contribute to neonatal mortality. The most severe lesions are considered critical congenital heart defects (CCHD). The specific definition of CCHD varies in the literature, but in general includes lesions that are dependent on early and prompt recognition to avoid patient harm.1 Prenatal detection of CCHD has been demonstrated to reduce neonatal morbidity and mortality, particularly in infants who are at high risk of rapid decompensation from lesions such as transposition of the great arteries or hypoplastic left heart syndrome.2-4 Prenatal detection leads to improved outcomes by allowing centers to anticipate the birth of a child with CCHD and plan accordingly. Also, prenatal detection affords the family the opportunity to make pregnancy related decisions such as termination.  Prenatal detection rates in the United States vary by lesion, but were 42% overall on a recent study, and slightly higher (50%) in neonates with CCHD.5 Thus there is considerable room for improvement of prenatal detection rates.  The population targeted in this metric is neonates ≤ 28 days of age undergoing surgical or catheter intervention for structural heart disease. This age range was chosen as it will include the most severe cases that are often considered CCHD. Improving detection rates in this population will likely have the most clinical impact of reducing perinatal morbidity and mortality. Note we do not propose this as a definition of CCHD, but rather use the term “severe structural CHD.”  Through use of this metric, centers will not only be able to track rates of prenatal detection of severe structural CHD, but also identify targets for improvement, such as certain lesion types and barriers to effective prenatal screening. It is anticipated that interventions meant to improve prenatal detection will cross disciplines to include all those involved in the care of pregnant women and their fetuses. | |
| **Clinical Recommendation(s)** | |
| ACC/AHA Guidelines:  Donofrio MT, Moon-Grady AJ, Hornberger LK, et al., Diagnosis and treatment of fetal cardiac disease: a scientific statement from the American Heart Association. Circulation. 2014;129:2183-2242.  Other guidelines:  Rychik J, Ayres N, Cuneo B, et al. American Society of Echocardiography guidelines and standards for performance of the fetal echocardiogram. J Am Soc Echocardiogr. 2004;17:803-810.  AIUM Practice Parameter for the performance of Obstetric Ultrasound Examinations - 2013 (<http://www.aium.org/resources/guidelines/obstetric.pdf>)  International Society of Ultrasound in Obstetrics and Gynecology, Carvalho JS, Allan LD, et al. ISUOG Practice Guidelines (updated): sonographic screening examination of the fetal heart. Ultrasound Obstet Gynecol. 2013;41:348-359. | |
| **Attribution** | |
| This measure should be reported by centers who wish to track the success of interventions aimed at improving prenatal detection. These interventions would likely involve engaging front line providers who screen for CHD in the community, including referring cardiologists, obstetricians, radiologists and maternal-fetal-medicine physicians. | |
| **Method of Reporting** | |
| Centers will perform a quarterly review of their institution’s surgical and catheter interventions for structural CHD in neonates ≤ 28 days of age. After exclusions as listed appendix, they will be left with the denominator. They will then review records to determine how many of those neonates had a prenatal diagnosis. This will be the numerator. | |
| **Challenges to Implementation** | |
| Labor intensive to review surgical and catheterization cases. Difficulty linking child records to maternal records. Lack of documentation and details of prenatal diagnosis. | |
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**References**

1. Slodki M, et al. Fetal cardiology: changing the definition of critical heart disease in the newborn. J Perinatol. 2016;36:575-80.
2. Bonnet D, et al. Detection of transposition of the great arteries in fetuses reduces neonatal morbidity and mortality. Circulation. 1999;99:916-8.
3. Tworetzky W, et al. Improved surgical outcome after fetal diagnosis of hypoplastic left heart syndrome. Circulation. 2001;103:1269-73
4. Morris SA, et al. Prenatal diagnosis, birth location, surgical center, and neonatal mortality in infants with hypoplastic left heart syndrome. Circulation. 2014;129:285-92
5. Quartermain MD, et al. Variation in Prenatal Diagnosis of Congenital Heart Disease in Infants. Pediatrics. 2015;136:e378-85

**APPENDIX**

**Exclusions**: (not to be included in denominator)

Non –structural lesion (e.g. cardiomyopathy for ECMO, VAD or heart transplant)

Specific structural lesions that are not amenable to prenatal detection: Anomalous coronary artery origin (e.g. ALCAPA), PDA.

**Definitions/Clarifications**:

A prenatal diagnosis is made if a prenatal examination by any provider (cardiologist, maternal-fetal-medicine, radiologist) detects structural heart disease that will either definitely or possibly require surgical intervention at any time in a child’s life

The prenatal diagnosis need not be accurate. For example, a prenatal diagnosis of double outlet right ventricle that turns out to be truncus arteriosus will still be included in the numerator.

**Limitations:**

It is recognized that this metric will not detect missed prenatal diagnoses for lesions that are operated on after 28 days of life (e.g. atrioventricular canal defects, tetralogy of Fallot, ventricular septal defects). The age limit of 28 days was chosen because these are the patients in whom a prenatal diagnosis is critical in order to prevent morbidity/mortality (e.g. hypoplastic left heart syndrome, transposition). Also, practically, it would be harder to determine whether a prenatal diagnosis was made for patients presenting for surgical repair later in childhood. This information is usually more readily available in neonates.

The age limit of intervention at ≤ 28 days’ gestation will exclude infants born prematurely who may wait longer than that time for intervention, as well as infants who suffer neonatal morbidity who wait longer than that time to recover before intervention. Infants who die prior to intervention or who are not candidates due to neonatal morbidities will also not be included.

**Worksheet:**

Centers will only report numerator/denominator, but a worksheet with additional clinical information can be filled out for missed cases of CHD that can help centers identify areas for improvement. (e.g. anatomy of lesion, whether prenatal screening was performed and by whom, maternal or other issues that may complicate screening: access to care, poor acoustic windows).