MINNESOTA NEONATAL PHYSICIANS P.A.
www.mnneo.org

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Congenital Pulmonary Airway Malformations (CCAM, CPAM, BPS)

I. **Introduction**

1. Congenital Pulmonary Airway Malformations occur in approximately 1 in 2000 live births but are increasingly being diagnosed prenatally due to increased utilization and accuracy of prenatal ultrasound. These lesions can usually be followed conservatively prior to birth, but occasionally require prenatal intervention and treatment. Prenatal care should focus on proper consultation for the parents and proper surveillance for the fetus.

II. **Definitions**

A. CPAMs are made up of a cluster of malformations previously known as congenital cystic adenomatoid malformations (CCAM), bronchopulmonary sequestrations (BPS) and congenital lobar emphysema. Differential diagnosis includes congenital diaphragmatic hernia, mediastinal teratoma, neuroblastoma, and forgut duplications cysts.

B. Various institutions have classified these lesions using different criteria. Original classification was done by Stocker with lesions grouped into three types; type I involving macrocystic lesions (>5mm diameter by ultrasound), type II (mixed macrocystic and microcystic), and type III (microcystic). Subsequent studies have included either microcystic or macrocystic subtypes, with mixed lesions placed in the macrocystic category.

III. **Preconceptual care**

1. Patients with prior children with CPAM are not felt to be at increased risk in future pregnancies.

IV. **Antenatal Management**

1. **Initial Encounter**

Level II Ultrasound

Assessment of size and character of chest mass including CVR measurement

Fetal Echo

Parents/mother given information sheet (from MNP website)

Introduce Perinatal/Neonatal Navigators (or make referral)

Tour Birth Center/Care Planning

V. **Perinatal Care Navigators**

1. Perform Genetics Screening Questionnaire for Pleuropulmonary Blastoma:

Family Questions: Has any brother, sister, parent, relative had any of the following:

Pneumothorax? _____
Kidney cysts? _____ (esp. cystic nephroma)
Ovarian Tumors; testicular tumors? _____ (esp. Sertoli-Leydig ovarian tumors; testicular seminomas, ovarian dygerminomas)
Lung cysts? _____
Thyroid nodules or **tumors**? _____
Eye or nasal tumors? _____
Childhood Cancers? _____

VI. **Consults to be scheduled**:

A. Neonatology
B. Pediatric Surgery
C. Genetics
   1. Referral to genetic counselor if any of above screening questions are positive. Screen for risk factors for Pleuropulmonary Blastoma.
   2. Families identified as high risk will be seen by Heidi Thorson MFM. She will review medical histories with parents and make referrals to PPB experts.
   3. Families identified as "high risk" for PPB will be seen prenatally and again postnatally by Drs. Messinger or Schultz as needed and followed by PPB protocols. Genetic counselors at Children's will be involved per Drs. Messinger or Schultz discretion.

   Genetic testing as indicated based on additional anomalies, family history, etc.
D. Tours of NICU and SCN
E. Fax updated information to designated NICU and NICU Office. NICU fax #:MSP (612) 813-6949, St. Paul (651) 220-7085. NICU office fax #: MSP (612) 813-5910, St. Paul (651) 220-7777

Mother/parents offered repeat consultation if hydrops or cardiac deviation develops (same neonatologist or surgeon)

VII. **Subsequent Perinatal Visits**

A. Start Antenatal testing at about 32 weeks gestation with weekly BPP/NST
B. Monitor fetal growth every four weeks
C. Monitor for hydrops weekly if CVR >1.4, or macrocystic CPAM
   i. If microcystic and CVR < 1.4 at < 32 weeks gestation, re-evaluate every 2 weeks
D. Consider steroids for patients with CVR ≥ 1.6 (Possible RCT at UCSF)

If macrocystic with significant mediastinal shift or hydrops, or microcystic with hydrops, notify Care Navigators for presentation for consideration of prenatal intervention.
If persistent large mass into third trimester with mediastinal shift or hydrops, notify Care Navigators for possible changes in delivery plans (EXIT, ECMO, Resection).

Transfer prenatal care to MPP by 34 weeks for planned delivery at tertiary center. Delivery at referring location not recommended due to 35% incidence of respiratory symptoms at birth (Children's Minneapolis data) but can be considered if ***ALL*** apply:

1. CPAM resolving over time
2. No other anomalies
3. Level II Nursery facility available at referring location
4. Parents educated on variability of clinical presentation
5. Potential for transfer to Children's Hospitals after delivery without guarantee of maternal transport.

VIII. Intrapartum Care

A. Timing of delivery

1. Delivery at 40-41 weeks (Ideal)

B. Route of delivery

1. Based on obstetrical indications, no benefit to cesarean section
2. Exception is for large masses late in pregnancy that may benefit from EXIT to ECMO or EXIT to Resection procedures

IX. Resuscitation Room

A. Routine resuscitation with intubation as indicated based on presentation
B. Place UAC and UVC only as indicated

X. NICU/SCN Care

A. CXR on admission
B. ABG or CBG if any respiratory symptoms
C. Pulse oximetry monitoring a minimum of 24 hours
D. Antibiotics if indicated based on risk factors and presentation
E. Can begin breast feeding ALD if asymptomatic and in RA
F. PIV and IVF with symptoms or if clinical course dictates
G. Consult Surgery and request original consulting surgeon to evaluate if possible
H. On-call surgeon to evaluate if infant symptomatic or primary surgeon not available

XI. **Postpartum Care**

A. Routine

XII. **Key Contacts**

Minneapolis Perinatal Clinic: (612) 863-4502,Fetal Therapy Coordinator: (612) 654-1602, NICU Office/Neonatal Consult Scheduling: (612) 813-6288,
Pediatric Surgeons: (612) 813- 8500

Saint Paul Perinatal Clinic: (651) 241-6270, Perinatal Navigators: (651) 241-6332,Fetal Therapy Coordinator: (612) 654-1602, NICU Office/Neonatal Consult Scheduling: (651) 220-6260, Pediatric Surgeons: (612) 813-8500