



Topic Expert Group: Medical care and clinical practice

Management of persistent pulmonary hypertension of the newborn infant (PPHN)

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Target group

Term and near-term infants >34 weeks of gestational age and parents

User group

Healthcare professionals, perinatal and neonatal units, hospitals, and health services

Statement of standard

Management of newborn infants with persistent pulmonary hypertension (PPHN) in a specialised centre improves mortality and morbidity.

Rationale

Persistent pulmonary hypertension of the newborn (PPHN) is characterised by sustained elevation of pulmonary vascular resistance (PVR) after birth resulting in extrapulmonary shunting from right to left via the fetal circulatory pathways (patent ductus arteriosus and patent foramen ovale). (1) PHN leads to severe hypoxaemia that may not respond to conventional respiratory support and to avoid severe cardiorespiratory failure. (2) The management of delivery and neonatal care should be transferred to a specialised centre to ensure optimal outcomes. (3,4)

Benefits

Short-term benefits

- Reduced mortality and morbidity (2,4,5)
- Reduced need for extra-corporeal membrane oxygenation (ECMO) (6)

Long-term benefits

- Reduced long-term morbidity (neurodevelopmental and cardiopulmonary outcome) (7)

Components of the standard

Component	Grading of evidence	Indicator of meeting the standard
For parents and family		
1. Parents are informed by healthcare professionals about persistent pulmonary hypertension (PPHN) in the newborn infant, treatment strategies as well as short- and long-term consequences.	B (High quality)	Patient information sheet



For healthcare professionals

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|--|--------------------------------------|------------------------|
| 2. A unit guideline on indications of pre- or postnatal transfer to a specialised centre, which may include facilities for ECMO, is adhered to by all healthcare professionals. (4,5) (see TEG Birth & transfer) | A (High quality)
B (High quality) | Guideline |
| 3. Training on the management of PPHN is attended by all responsible healthcare professionals. | B (High quality) | Training documentation |

For perinatal and neonatal unit

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|--|--------------------------------------|-----------|
| 4. A unit guideline on indications of pre- or postnatal transfer to a specialised centre, which may include facilities for ECMO is available and regularly updated, to include: (4,5) <ul style="list-style-type: none">• Resuscitation and early management are focused on optimal lung recruitment and ventilation, and based on neonatal resuscitation guidelines. (2)• Diagnosis is confirmed by detection of pre- and post-ductal saturation difference of 5-10%.• 2D echocardiography is performed to rule out the presence of cardiac anomalies and assess right heart function; the degree of pulmonary hypertension is classified relative to systemic blood pressure. (5)• Inotropic drugs are used to support organ perfusion. (5)• In case of poor organ perfusion, treatment with inhaled nitric oxide (iNO) is started. (5,7,8)• If there is insufficient response to iNO despite optimal lung recruitment, i.v. drugs such as phosphodiesterase inhibitors (sildenafil, milrinone) or prostaglandin are considered. (3)• ECMO is considered according the extracorporeal life support organisation (ELSO) guidelines (OI>20). (4)• Local protocols are updated in regular intervals to ensure individualised therapy. | A (High quality)
B (High quality) | Guideline |
|--|--------------------------------------|-----------|



For hospital		
5. Training on the management of PPHN is ensured.	B (High quality)	Training documentation
6. Access to 24/7 echocardiography, radiology, and laboratory support is ensured.	B (High quality)	Guideline
7. ECMO is only provided in designated centres.	B (High quality)	Guideline
For health service		
8. A national guideline on indications of pre- or postnatal transfer to a specialised centre, which may include facilities for ECMO, is available and regularly updated. (see TEG Birth & transfer)	B (High quality)	Guideline

Where to go – further development of care

Further development	Grading of evidence
For parents and family N/A	
For healthcare professionals N/A	
For perinatal and neonatal unit N/A	
For hospital N/A	
For health service	
<ul style="list-style-type: none"> Develop research into optimal saturation targets, use of inodilators and additional drugs to treat persistent pulmonary hypertension (PPHN). 	A (Low quality)
<ul style="list-style-type: none"> Provide long-term multidisciplinary standardised follow-up after discharge. (7) 	A (Moderate quality)
<ul style="list-style-type: none"> Set up an European-wide database for newborn infants with PPHN. 	A (Low quality)

Getting started

Initial steps
For parents and family
<ul style="list-style-type: none"> Parents and family are verbally informed by healthcare professionals about persistent pulmonary hypertension (PPHN), treatment strategies as well as short- and long-term consequences.



For healthcare professionals

- Attend training on management of PPHN.
- Raise awareness to diagnose PPHN.

For perinatal and neonatal unit

- Develop and implement a unit guideline on indications of pre- or postnatal transfer to a specialised centre, which may include facilities for ECMO.
- Develop information material about PPHN, treatment strategies as well as short- and long-term consequences for parents.

For hospital

- Support healthcare professionals to participate in training on management of PPHN and the use of licensed drugs.
- Ensure facilities and devices for optimal care and follow-up care.

For health service

- Develop and implement a national guideline on indications of pre- or postnatal transfer to a specialised centre, which may include facilities for ECMO.

Source

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6. The Neonatal Inhaled Nitric Oxide Study Group. Inhaled nitric oxide in full-term and nearly full-term infants with hypoxic respiratory failure. *N Engl J Med*. 1997;336(9):597–604.
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Lifecycle

5 years/next revision: 2023

Recommended citation



european standards of
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