Non-Immune Fetal Hydrops (NIHF) MATY090	
Type: Guideline	HDSS Certification Standard:
Issued by: Maternity PPG Group	**Version:** 1
Applicable to: Maternity, Childrens, Radiology	**Contact person:** O & G SMO
Lead DHB: HVDHB	**Level:**

Hutt Maternity Policies provide guidance for the midwives and medical staff working in Hutt Maternity Services. Please discuss policies relevant to your care with your Lead Maternity Carer.

Purpose:

To provide guidance and a consistent approach for the accurate diagnosis and management of people and babies presenting with non-immune fetal hydrops in the Secondary Care setting.

Scope:

For the purposes of this document, staff will refer to:

All staff within Hutt Valley DHB. This includes staff not working in direct contact with patients/consumers. Staff are taken to include anyone engaged in working to the Hutt Valley DHB. This may include but is not limited to:

- Employees irrespective of their length of service
- Agency workers
- Self-employed workers
- Consultants
- Third party service providers, and any other individual or suppliers working in Hutt Maternity, including Lead Maternity Carers, personnel affiliated with third parties, contractors, temporary workers and volunteers
- Students

Definitions:

- **SDP** Single deepest pool
- **SVT** Supraventricular tachycardia
- **SLE** Systemic lupus erythematosus
- **TTTS** Twin-to twin-transfusion syndrome
- **CPAM** Congenital pulmonary airway malformation
- **G6PD** Glucose -6-phosphate dehydrogenase
- **MCA** Middle cerebral artery
- **PSV** Peak systolic velocity
- **PTL** Preterm labour
- **TOP** Termination of pregnancy

Background

Non-immune hydrops fetalis / fetal hydrops is now responsible for 90% of all cases of fetal hydrops.

Estimated incidence of 1:1500 to 1:3800 births.

Pathogenesis not clear, but associated with numerous potential mechanisms and underlying disorders.

NIFH has a poor prognosis for the fetus, but can also have consequences for the pregnant person e.g. 10% incidence of Mirror syndrome.

Diagnosis

NIFH is the presence of any **TWO** of the following ultrasound findings given below:

- Ascites
- Pleural effusion (any fluid significant)
- Pericardial effusion: > 2mm significant
- Skin oedema: > 5mm on chest and scalp
- Polyhydramnios: SDP > 8cm and/or AFI > 24cm
- Placentomegaly: > 4cm thickness

Differential Diagnosis

- Immune hydrops
- Isolated fluid collection
    - Ascites
    - Pleural effusion
    - Pericardial effusion
- Skin oedema
- Polyhydramnios or placentomegaly

Causes

Aneuploidy	• **Most common cause of NIFH under 24 weeks: trisomy 21 Down's syndrome, 45XO Turner's syndrome** • **Genetic: Noonan's syndrome, Cornelia de Lange syndrome**
Cardiovascular anomalies	• Structural: most common cause of NIFH over 24 weeks • Functional: transposition of great vessels, Ebstein's anomaly, truncus arteriosus • Arrhythmias: tachyarrhythmia (SVT), bradyarrhythmia (complete heart block e.g. due to maternal SLE)
Vascular	• Shunt • Thrombosis
Thoracic abnormalities	• CPAM • Congenital diaphragmatic hernia • Masses • Pulmonary sequestration

	• Chylothorax, hydrothorax • Airway obstruction
Non-cardiac/thoracic anomalies	• Lymphatic • Gastrointestinal • Genitourinary • Neurological / decreased movement
Fetal anaemia	• Decreased production: Parvovirus B19, infiltration / storage diseases, myeloproliferative / congenital leukaemia • Increased loss: intrinsic cell abnormality (α thalassaemia, G6PD) • Haemangioma • Fetomaternal haemorrhage (e.g. abruption) • Fetal haemorrhage (intraabdominal, intracranial)
Infectious disease	• Parvovirus • Toxoplasmosis • Syphilis • Varicella • Adenovirus • Coxsackie • CMV • Herpes • Listeria
Metabolic storage disease	• Mucopolysaccharidoses
Placental	• TTTS / TRAP sequence • Trauma • Cord anomalies • Chorioangioma
Tumours	• Fetal sacrococcygeal teratoma
Idiopathic	

Clinical Presentation

- 35% cases detected incidentally
- Symphyseal-fundal height palpating large for dates
- Decreased fetal movement
- Mirror syndrome in pregnant person

Important History

- Personal and family history of inheritable disorders associated with:
    - α Thalassaemia
    - Metabolic syndromes
    - Genetic syndromes

- Infectious exposures – especially Parvovirus B19
- Consanguinity

Investigations

Investigations of the pregnant person

- Baseline blood pressure and urinalysis (looking for proteinuria)
- Full blood count
- Blood group and red blood cell antibody screen
- Hb electrophoresis (Thalassemia)
- Kleihauer Betke test (fetomaternal haemorrhage)
- Serology: parvovirus B19, toxoplasmosis, rubella, syphilis
- Herpes simplex virus (HSV) if recent primary infection
- Auto-antibodies: anti-SSa(Ro) and anti-SSb(La) if SLE

Fetal investigations

When NIFH is detected on ultrasound the following should also be reviewed; if not, then a subsequent scan ought to be requested

- Detailed anatomy review (tertiary scan)
- MCA PSV
- Echocardiogram
- Amniocentesis for karyotype, PCR for possible TORCH pathogens, storage in case further testing subsequently required

⚠ **Urgent referral to Wellington Maternal Fetal Medicine service needs to be made by the Obstetric team (after consultation with Obstetric SMO on-call)**

Fetal anaemia

- MCA PSV >1.5 MoM for gestation is suggestive of moderate to severe fetal anaemia (Hb < 80)

Management

Management will depend upon the underlying cause and the level of fetal anaemia

- Early diagnosis: 50% association with chromosomal problems and thus a poor prognosis → offer TOP
- Selective therapeutic intervention
- SVT: Maternal / fetal administration of digoxin ± anti-arrhythmic agents
- Complete heart block: maternal administration of steroids
- Fetal anaemia: cordocentesis and intrauterine blood transfusion
- Pleural effusion alone: pleuroamniotic shunting

Prognosis

In hydrops, the fetal prognosis depends upon:

- Gestation (earlier diagnosis has worse prognosis)
- Pleural effusions (worse prognosis, especially if under 20 weeks)
- Underlying aetiology

In an ongoing pregnancy:

- 10% develop PET (mirror/Ballantyne syndrome)
- Risk of polyhydramnios / PTL / abruption – amnioreduction may be required
- Mode of delivery uncertain
- Neonatal palliation
- Overall perinatal mortality 50-98%

There is a low risk of recurrence in a future pregnancy if no inheritable disorder is identified.

Mirror Syndrome

Mirror syndrome is a rare association of fetal and placental hydrops in association with pre-eclampsia. It is sometimes known as 'triple oedema' as it affects the fetus, placenta and pregnant person.

The name 'mirror syndrome' refers to the similarity between maternal oedema and fetal hydrops.

It can be confused for pre-eclampsia.

Mirror syndrome tends to resolve as the fetal hydrops resolves and/or after birth.

References:

- Hyett J .Fetal Hydrops.In: Fetal Medicine:Basic Science and clinical practice. Rodeck CH, Whittle MJ, editors. London: Churchill Livingstone, 2008.
- New Zealand Maternal Fetal Medicine Network. 2012. Nonimmune Hydrops Fetalis. doi

Keywords for searching:

1. Fetal Hydrops

2. NIFH

3. Non-immune

4. Mirror Syndrome

5. Pregnancy

Informed Consent

The right of a consumer to make an informed choice and give informed consent, including the right to refuse medical treatment, is enshrined in law and in the Code of Health and Disability Consumers' Rights in New Zealand. This means that a woman can choose to decline treatment, referral to another

practitioner, or transfer of clinical responsibility. If this occurs follow the process map on page 18 of the Referral Guidelines (Ministry of Health, 2012).

Tangata Whenua Statement

The Women's Health Service recognises the rights and responsibilities of Māori as tangata whenua and Treaty Partners. This allows and acknowledges the importance of cultural diversity in all aspects of our care and practice in Aotearoa New Zealand.

As stated in Te Pae Amorangi (Hutt Valley DHB Māori Health Strategy) 2018-2027, Hutt DHB as a Crown agency is committed to our role in maintaining active relationships with iwi, under Te Tiriti o Waitangi. This strategy recognises the established principles of Partnership, Participation and Protection and recognises steps towards the reviewed interpretation of Te Tiriti principles to date (from the Wai 2575 claim into health). These are tino rangatiratanga, equity, active protection, partnership and options.

Attention in particular is drawn to:

- **Article one – Kāwanatanga**: actively engaging and working alongside with local iwi through the Hutt Valley Māori Health Unit
- **Article two – Tino Rangatiratanga**: Self-autonomy, self-determination; the responsibility to enable Māori to exercise their authority over their own health, determinants and definition of health
- **Article three – Ōritetanga**: equal health outcomes of peoples; ensuring that policy, guidelines or programmes do not further perpetuate any inequity
- **Article four (the 'oral clause') – Wairuatanga**: spirituality; thriving as Māori and the importance of health providers understanding health in te ao Māori (the Māori world), acknowledging the interconnectedness and inter-relationship of all living and non-living things.