

Floppy Infant - Full Clinical Neonatal Guideline – Joint Derby and Burton

Reference no.: NIC NE 05/ June 22/v003

1. Introduction

To ensure a standardised approach to the management of "floppy infants"

2. Aim and Purpose

To facilitate staff in making a timely diagnosis and management plan

3. Definitions

"Floppy" is used to describe a baby that has a subjective decrease in resistance to passive movement. This can be due to problem at any level of the neuromuscular system.

It is important to remember that hypotonia can be present in sepsis, premature babies and babies with hypoglycaemia.

4. Abbreviations

CK Creatinine kinase

CMD Congenital muscular dystrophy

CSF Cerebrospinal fluid

CT Computed tomography

DNA Deoxyribonucleic acid

EEG Electroencephalography

EMG Electromyelography

MRI magnetic resonance imaging

NCS Nerve conduction studies

TFTs Thyroid function tests

PWS Prader-Willi syndrome

SMN Survival motor neurone

VLCFAs Very long chain fatty acids.

5. Main body of Guidelines

History

A thorough and careful history is important to guide initial investigations. Questions that should be asked include:

Maternal medical history: Diabetes

Myotonic dystrophy

Neuromuscular disease

Muscle weakness

 Any significant family history: Affected siblings or parents Consanguinity

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Previous unexplained neonatal/infant death

Pregnancy: Drug exposure

Reduced fetal movements,

Polyhydramnios/oligohydramnios,

Resuscitation at birth: Apgar scores

Umbilical cord gases

History since delivery: Respiratory effort

Feeding difficulty

Alertness

Seizures/abnormal movements

Spontaneous activity

Cry

Examination

Examination should firstly confirm the presence of hypotonia in an infant. There are a number of manoeuvres to elicit reduced tone. These include

- Head lag
- "Rag-doll" posture on ventral suspension
- Observation of a "frog-leg" posture
- Vertical suspension
- "Scarf" sign positive

Once hypotonia has been confirmed the next step is to determine whether the problem is of central or peripheral origin. Typical features of each are shown in the table below.

Central hypotonia	Peripheral hypotonia
Normal movement	Reduced or absent spontaneous anti- gravity movement
Normal or brisk tendon reflexes	Normal or reduced reflexes
	Muscle fasciculation (rare but diagnostic)

Additional clinical features that may indicate a specific diagnosis include:

- Hepatosplenomegaly storage disorders, congenital infection
- Hypopigmentation, undescended testes Prader Willi
- Abnormal odour metabolic disease

Check if there are any maternal features to suggest myotonic dystrophy

Diagnostic approach

Once a decision about a central or peripheral aetiology has been made, investigations should be directed accordingly to identify the cause of the hypotonia, as outlined below.

APPROACH TO THE "FLOPPY" INFANT "FLOPPY INFANT" DETAILED HISTORY **EXAMINATION** CLINICAL FEATURES Normal/† reflexes Absent/ reflexes Normal movement Weakness Dysmorphism Seizures Muscle wasting Reduced Contractures consciousness Arthrogryposis Karyotype CENTRAL PERIPHERAL Genetics review Blood gas Consider: First-line investigations Ammonia Cranial ultrasound DNA for SMN deletion Lactate CT MRI DNA for CMD Plasma amino acids EEG CSF lactate Renal cysts (seen in glycine Zellweger's syndrome) serine Urine amino/organic acids Second-line investigations Acylcarnitine NCS/EMG **VLCFAs** Muscle biopsy **TFTs** Neostigmine test DNA for PWS NB These should be discussed with the paediatric neurologist and are NB-has sepsis been usually done at the tertiary centre considered and excluded?

Remember that premature babies and babies with sepsis, hypoglycaemia and jaundice can also present with hypotonia

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6. References (including any links to NICE Guidance etc.)

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- Paro-Panjan D and Neubauer D. Congenital hypotonia: is there and algorithm. Journal of child neurology. 2004; 19: 439-442.
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 Brain and development. 2003; 27: 457-476.
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7. Documentation Controls

Reference Number	Version:		Status				
NIC NE 05/ June 22/v003	3		Final				
Version /	Version	Date	Author	Rea	Reason		
Amendment History	V003	June 2022		Guideline required to be made cross site with Burton			
Intended Recipients: Neonatal and Paediatric Consultants							
Training and Dissemination:							
Development of Guideline: Dr Gitika Joshi Job Title: Neonatal Consultant							
Consultation with: All NICU and Paediatric Consultants							
Linked Documents: None							
Keywords: Hypotoxic Baby, Floppy Infant							
Business Unit Sign Off			Group: Paediatric Guidelines Group Date: 08/06/2022				
Divisional Sign Off			Group: Women's and Children's Clinical				
		Governance Group Date: 28/06/2022					
			July 2022				
Review Date			June 2025				
Contact for Review			Dr Gitika Joshi				