Management and Investigation of Sudden Unexpected Death or Collapse in Apparently Healthy Term Infants

Definition
The Sudden Unexpected Postnatal Collapse of apparently healthy term infants includes any baby at or near term (>35 weeks gestation) who
• Is well at birth – deemed well enough to have routine postnatal care
• Collapses unexpectedly – is in a state of cardiorespiratory extremis such that resuscitation with intermittent positive pressure ventilation is required
• Either dies or goes on to require intensive care

Incidence
The incidence of sudden unexpected and unexplained death or neonatal collapse is reported as between 0.035/1000 to 0.4/1000 live births. Although rare, greater than half of these infants die and the majority of survivors have significant long term neurodevelopmental morbidities. 1 2 3 4 5 6 7

Background
It is clear from the retrospective examination of neonatal sudden unexplained death in infants (SUDI) in NSW8, that the history, examination and investigation of these cases are not always completely adequate. Of the 123 neonatal sudden unexplained infant deaths reported (1996-2008), 37 (30%) occurred in the first 7 days of life8.

The British Paediatric Surveillance Unit performed a recent study looking at the sudden unexplained collapse or death of apparently healthy term infants in the first 12 hours of life. In this study, 30 out of 45 cases had no identifiable underlying disease/abnormality but in 24 there was clinical or pathological evidence of airway obstruction9. Ten out of the twelve infants who died had a post-mortem and a cause of death was identified in 50 % of cases. Underlying conditions including infection, congenital cardiac defects, metabolic defects, intracranial haemorrhage/infarction, meconium aspiration syndrome, severe chronic anaemia (related to parvovirus infection), congenital diaphragmatic hernia, central hypoventilation syndrome and adrenal hypoplasia were all identified as causative in infants.10

In infants who die in the first week of life, more than 50% are explained after post-mortem examination. The information obtained from investigations and from post-mortem examination is extremely important for the management and prognosis of infants who survive such an event, for counselling of families and for potential future pregnancies.

Neonatal unexplained death is a rare condition with the above important implications for the baby and family. For these reasons the following has been developed:
• A protocol with checklists for history, examination and investigations
• SUDI and near miss SUDI within 7 days of birth are now listed by the
  Australian Paediatric Surveillance Unit (APSU) for detailed surveillance

A reminder that such cases by law must be referred to the Coroner; however, a
discussion with the forensic pathologist can enable a paediatric perinatal post mortem.
The pathological investigation of infants dying in the first week of life has a higher
diagnostic yield compared with later SUDI.¹¹

Risk Factors

In the reported literature, many of these infants are found face down on the mother’s
breast suggesting that airway compromise may be a contributing factor. The
association of skin-to-skin, breast feeding and prone position have been well
documented.² Other risk factors include primaparous women, recent maternal
analgesia, bed-sharing and unattended mothers.¹⁰ In the study by Polberger et al all
deaths or collapse occurred between 11pm and 6am while six out of ten infants in the
study by Burchfield et al collapsed in the early morning hours.¹ The history of the
event occurring in the early hours of the morning was not consistent in all the reported
literature.

Approach to Sudden Unexpected Death or Collapse

- History

All infants who present with sudden unexpected collapse or death in the neonatal
period need a thorough history and clinical examination to in an attempt to establish
the cause for the event. If at all possible, this history and examination should be
performed by a neonatologist/ paediatrician or the most senior neonatal/paediatric
clinician.

Maternal history
- Maternal antenatal care
- Maternal gravidity
- Maternal infection - GBS status, urinary tract infections, fever during labour
- Previous pregnancy resulting in stillbirth, neonatal death or collapse
- Maternal complications during pregnancy
- Maternal medications or substance use during pregnancy
- Analgesia during labour
- Post delivery analgesia including timing
- Family history – cardiac disease, genetic conditions, consanguinity, metabolic
disease
- Parents’ occupations

The delivery
- Mode of delivery
- Onset of labour
- Presence of meconium stained liquor
- Resuscitation required at birth including Apgar scores
- Arterial cord gas if available

The presentation
- Who found the baby?
- Who was in the room at the time of event?
- Time of the event
- Position the infant was last placed
- Position and place the infant was found
- Any items near infants face – toys, bumper pads, positional supports
- Was the infant using a dummy at the time of the collapse
- When last reviewed was the baby feeding, awake but not feeding or asleep
- Date and time of last observation/check before being found
- Feeding – breast, formula or both. Timing of last feed.
- Infant medications
- In the 24 hours prior to the collapse did the infant have an acute illness
- In the 24 hours prior to the collapse was there a history of trauma/fall

A detailed questionnaire can be found on the APSU website (www.apsu.org.au)

If the collapse occurs in the delivery suite, it may be helpful to obtain photos’ of the baby, with or without the mother, from the family in the time leading up to the collapse. This may be especially useful if the collapse is thought to be due to positioning of the baby.

**Examination**
The examination should be thorough and systematic as detailed in the PSANZ perinatal audit guidelines. Anthropometric data should be documented (weight, length and head circumference) and percentiles charted on the WHO growth charts. Any obvious dysmorphic features should be highlighted. Clinical photographs (with consent) should be considered if it is unlikely that the baby will be reviewed by a geneticist.

**Suggested Investigations on infants presenting with sudden unexpected collapse**
NEONATAL COLLAPSE

↓

HISTORY

↓

CLINICAL EXAMINATION

↓

PRIMARY INVESTIGATIONS

- **SEPSIS**
  - Blood – FBC, Coagulation studies, CRP, cultures, parvovirus PCR
  - Urine – bacterial culture, CMV PCR
  - CSF – bacterial culture, HSV and enterovirus PCR
  - Stool – enterovirus
  - NPA – viruses
  - CXR
  - *See below for maternal investigations relevant to sepsis*

- **CARDIAC DISEASE**
  - ECG – conduction defects
  - CXR
  - Cardiac ultrasound

- **METABOLIC DISEASE**
  - Blood – venous gas, electrolytes, CMP, ammonia, LFT, glucose, amino acids, organic acids, lactate, uric acid, acyl carnitine, sulphocysteine
  - Urine – metabolic screen
  - CSF – lactate and pyruvate
Suggested Investigations on infants presenting with sudden unexpected death

<table>
<thead>
<tr>
<th>Investigation</th>
<th>Clinical photographs* 12,13</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>AP view – whole body including limbs</td>
</tr>
<tr>
<td></td>
<td>PA view</td>
</tr>
<tr>
<td></td>
<td>Lateral view of the body</td>
</tr>
<tr>
<td></td>
<td>Lateral view of the face</td>
</tr>
<tr>
<td></td>
<td>Frontal view of face</td>
</tr>
<tr>
<td></td>
<td>Photograph of any detected abnormality</td>
</tr>
<tr>
<td>Blood investigations</td>
<td></td>
</tr>
<tr>
<td>-------------------------------------------------</td>
<td></td>
</tr>
<tr>
<td>Blood culture (cardiac tap)</td>
<td></td>
</tr>
<tr>
<td>Dried Blood spot (NBST)</td>
<td></td>
</tr>
<tr>
<td>Sepsis</td>
<td></td>
</tr>
<tr>
<td>Metabolic Disease</td>
<td></td>
</tr>
<tr>
<td>DNA storage</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Skin and muscle</th>
</tr>
</thead>
<tbody>
<tr>
<td>Skin Surface Swabs for bacterial culture</td>
</tr>
<tr>
<td>Skin and muscle biopsy</td>
</tr>
<tr>
<td>Mitochondrial disorder</td>
</tr>
<tr>
<td>Skin biopsy for fibroblast line as a source of DNA storage</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Imaging</th>
</tr>
</thead>
<tbody>
<tr>
<td>CXR, Skeletal survey</td>
</tr>
<tr>
<td>Congenital anomalies</td>
</tr>
<tr>
<td>Non Accidental Injury</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Post mortem</th>
</tr>
</thead>
<tbody>
<tr>
<td>Including anthropometric measurements (weight, length, head and abdominal circumferences).</td>
</tr>
<tr>
<td>Preferably performed by a paediatric or perinatal pathologist.</td>
</tr>
<tr>
<td></td>
</tr>
</tbody>
</table>

*The clinical photographs should be taken from above; the baby should be naked. Informed consent should be documented. Clinical photographs are particularly useful if babies display dysmorphic features and if a post mortem is not performed.

- **Suggested maternal investigations**

The following investigations are recommended for mothers whose newborn infant presents with unexpected collapse or death

<table>
<thead>
<tr>
<th>Placenta</th>
</tr>
</thead>
<tbody>
<tr>
<td>Macroscopic examination</td>
</tr>
<tr>
<td>Placental swabs for bacterial and viral culture (bacterial cultures should be taken between the chorion and amnion)</td>
</tr>
<tr>
<td>Histopathology</td>
</tr>
</tbody>
</table>
Follow-up of families

These families should be followed up by a neonatologist or paediatrician, a social worker and a geneticist if appropriate at 6-8 weeks after the death. Any outstanding results should be relayed to the family including the preliminary results of the post mortem as soon they become available.

References


Author: Dr Tracey Lutz (2013)
1.4 Clinical examination of baby checklist

Please tick appropriate box and complete details as required

Baby measurements
1. Crown - heel (stretched) cm
2. Head circumference cm
3. Weight g

If stillborn
Estimated date of IUDF
Maceration degree
Fresh, no skin peeling
Mild skin peeling
Moderate, much skin peeling but no secondary comprehensive changes or decomposition
Marked, advanced

HEAD AND FACE
Head
Relative normal
Anoplastic
Abnormal shape
If abnormally shaped, describe:

Eyes
Normal
Straight
Upstaring
Glaucoma normal
Eyes very small
Lens opacity
Eyelids fused
If other, describe:

Nose
Normal
Asymmetrical
Nasal pits
Apparent patent
Single nostril
If other, describe:

Mouth
Normal size
Upper lip intact
Left
Bilateral
Palate intact
Mandible normal
If other, describe:

Ears
Normal
Lowest
Other
If other, describe:

NECK
Normal
Mats
Describe:

CHEST
Normal
Long & narrow
Short & broad
If Spina bifida, describe:

ABDOMEN
Normal
Flattened
Distended
Omphalocele
Gastrostomy

BACK
Normal
Spina bifida
If Spina bifida, describe:

GENITALIA
Anus
Normal
Imperforate
Other
If other, describe:

Gender
Male
Female
Ambiguous
Other
If other, describe:

Penis
Normal
Very small
Hypospadias, level of opening
Scrotum
Normal
Abnormal
If abnormal, describe:

Testes
Descended
Undescended
Other
If other, describe:

FEET
Unilateral opening
Present
Absent/undetectable
Vaginal introitus
Present
Absent/undetectable
Cleft
Enterovesical or Other
If other, describe:

Revised gestational age
Based on

Examined by:
(Date)
Summary of key findings: