

**Guidelines for the Investigation of
Newborn Infants who suffer a
Sudden and Unexpected Postnatal Collapse
In the First Week of Life**

**Recommendations from a Professional Group on Sudden
Unexpected Postnatal Collapse**

March 2011

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Funded by WellChild



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Guidelines for the investigation of newborn infants who suffer a sudden and unexpected postnatal collapse in the first week of life

Summary of recommendations

- Infants who suffer a sudden and unexpected cardiorespiratory collapse within the first week of life should be recognised as having an increased risk of congenital anomaly or metabolic disease as an underlying cause for their collapse
- All infants who suffer a sudden and unexpected cardiorespiratory collapse within the first week of life should undergo comprehensive investigation to determine the underlying cause.
- Such an investigatory process will involve interdisciplinary liaison to maximise diagnostic yield whilst minimising unnecessary tests for the child.
- A detailed history of the family and situational events is essential and should be obtained by a senior member of medical staff
- All infants who die from such collapse should be notified to a Coroner/Procurator Fiscal
- All infants who die should undergo post mortem performed by a perinatal pathologist
- A detailed multiprofessional case review should follow the investigation of any unexpected infant death

Guidelines for the investigation of newborn infants who suffer a sudden and unexpected postnatal collapse in the first week of life

Introduction

This document is intended to provide guidance for the appropriate investigation of infants who suffer a sudden and unexpected collapse in the first week after birth. The guideline covers investigation of both survivors and those who die. It takes account of published guidance for the investigation of Sudden Unexpected Death in Infancy (SUDI)(1) and input from UK perinatal professionals.

Definition

For the purpose of these guidelines, an infant who suffers a 'Sudden Unexpected Postnatal Collapse' includes any term or near term (>35 weeks gestation) infant who

- is well at birth (normal 5 minute Apgar score and deemed well enough to have routine postnatal care) and,
- collapses unexpectedly ie discovered in a state of cardiorespiratory extremis such that resuscitation with intermittent positive pressure ventilation is required and,
- collapses within the first seven days of life and,
- who either dies or goes on to require intensive care or develops an encephalopathy

Target users

- All paediatricians involved in the care of the newborn
- Pathologists involved in perinatal post mortem
- Radiologists involved in imaging of the newborn

Purpose of guidelines

Sudden unexpected postnatal collapse (SUPC) in the days and hours after birth occurs in 0.03-0.08/1000 livebirths (2-13) with an incidence in the first twelve hours of 1 in 20,000 live births within the United Kingdom (14). There is significant mortality and long-term neuromorbidity associated with such collapse. The approach to investigation is currently highly disparate, the range of underlying aetiologies not well elucidated and the potential for treatment or genetic counselling limited.

Underlying congenital anomalies and conditions arising through pregnancy and birth are over-represented in this group of infants compared to older infants who suffer sudden unexpected death (15). The proximity to birth and extra-uterine adaptation necessitates a thorough examination of perinatal and situational factors. Although the published guidance for the investigation of Sudden Unexpected Death in Infancy (SUDI) should encompass all infant deaths, the emphasis is on those dying outside hospital after the first month of life, where parental lifestyle factors and non-accidental injury may have more contribution. In SUDI cases, which occur outside a hospital, there is rarely the opportunity to carry out investigations prior to death. For babies who collapse in hospital and who subsequently die, there is often the opportunity to carry out investigations during the period of intensive care. In addition the SUDI guidance does not include investigation of survivors who account for around 50% of those infants suffering SUPC.

This document is not intended to replace or duplicate the guidance for the investigation of Sudden Unexpected Death in Infancy (1) but aims to improve the likelihood of diagnosis in this group of newborn infants. This guideline does not supersede the national arrangements for a joint agency 'rapid response' to unexpected deaths in infancy and childhood (16) or for local risk management review for survivors.

The guideline intends to standardise the investigative approach to this group of infants in order to

1. establish the most likely cause of collapse
2. document any underlying disease whether or not considered a direct cause for collapse
3. provide information relevant to the future reproductive health of parents and the future health of siblings of the case infant.
4. collect and secure evidence where required by the Coroner/Procurator Fiscal
5. clarify prognosis and ongoing management in those who survive

The guideline does not intend to address professional responsibilities or recommend an approach to management of such infants; instead the emphasis is the optimisation of the investigatory process. Consideration of transfer for specialist management including cooling treatment should be addressed by local network guidelines.

Background

Sudden unexpected postnatal collapse in apparently well term babies, in the first week of life, as defined above, is a rare event. Although infrequent in any one centre these infants are well described in the literature and commonly suffer death or severe long-term neurological sequelae (2-13). A study conducted through the British Paediatric Surveillance Unit in 2009 estimated the UK incidence of SUPC in the first 12 hours as 0.05/1000 live births of whom 27% died (14). There is a highly disparate approach to investigation of these infants throughout the United Kingdom.

In order to protect parents from the perceived ordeal of post mortem clinicians may attribute death to the hypoxic-ischaemic sequelae without elucidating the cause of the initial collapse. In those infants where careful perinatal post mortem has been undertaken, valuable information has often been found. Underlying conditions such as antenatal brain injury, infection, metabolic defects, congenital central hypoventilation syndrome, congenital adrenal hypoplasia and cardiac abnormalities such as structural anomalies, cardiomyopathy, infarction and conduction abnormalities have all been described (6)(10)(15)(Appendix 4). Indeed, in those infants dying unexpectedly within the first week of life, almost 60% are explained following post mortem (15) compared to later SUDI deaths where a new but preceding diagnosis is confirmed in only 37% of cases (17), figure 1. When babies die, such pathologies have important implications for the parents, not only in explaining their child's catastrophic deterioration but also in aiding the grieving process and providing information for future pregnancies (11). In those infants who survive, a detailed set of investigations will optimise the chance of finding an explanation for the collapse which will have implications for management and prognosis of the infant.

Concern about this group of infants was raised in 2008 and UK neonatologists sought to study the incidence of SUPC through the British Paediatric Surveillance Study. This was preceded by an informal survey of members of the British Association of Perinatal Medicine, which showed that many neonatologists had experience of such infants although they were rare in any one centre. There was concern that the typical experience of the majority was unexpected and unpredictable deterioration in an apparently well infant with no apparent cause being found using standard investigations. Many neonatologists reported a poor uptake of post mortem examination and there was inconsistency of referral to the Coroner/Procurator Fiscal despite fulfilling the criteria for Sudden Unexpected Death in Infancy.

Infants suffering SUPC in the first week of life ought to undergo the process recommended by the Kennedy protocol, but there is a very strong case for additional data and investigations for those dying in the immediate postnatal period. In addition published literature suggests that around 50% of babies survive following SUPC and these would not be covered by the Kennedy protocol.

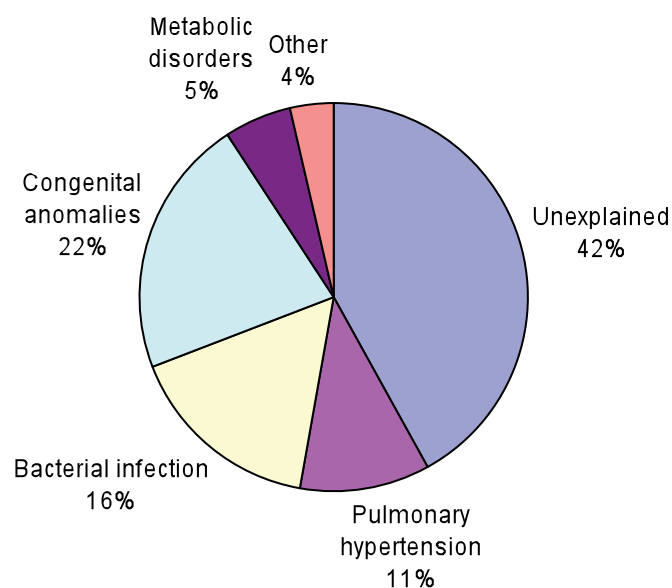


Figure 1. Causes of death from a series of 55 sudden unexpected neonatal deaths undergoing post mortem examination. Adapted from Weber et al (15)

At the end of December 2008 a group of perinatal professionals was assembled to seek consensus on guidelines for investigation of such infants. This group comprised neonatologists, biochemists, pathologists, radiologists and experts in SUDI, and included representation from obstetrics, midwifery and bereavement groups (Appendix 5).

A set of investigations has been agreed which the group believe will determine, as far as possible, the causes of SUPC in both those dying and in survivors. There is a strong recommendation for early involvement of the Coroner/ Procurator Fiscal and for the post mortem examination to be performed by a paediatric/perinatal pathologist. In addition, the group recommend retention of brain for optimal neuropathology as well as seeking consent for retention of tissue for recognised and unrecognised future purposes. A minimum dataset to aid clinicians in their investigation of such cases is included. The guidelines focus on an investigatory approach and do not extend to recommendations about process or case review following such events. It is expected that local procedures in investigating serious adverse events will take place and reference to the Multi-agency Protocol for Care and Investigation of Sudden Unexpected Death in Infancy is recommended (16).

Potential organisational barriers in applying the recommendations have been discussed by the group and by stakeholders (Appendix 6). Where barriers have been perceived, primarily in Coroner legislation, these have been resolved by comprehensive discussion with the Coroners' Society and the introduction of new legislation in 2011 which improves accessibility to perinatal post mortem for all infants. In addition the economic implications of a Coroner post mortem for all infants dying following SUPC has been explored with the Coroners' Society and decided that for individual Coroner districts this would not be excessive. The potential cost implications of applying the other recommendations have also been considered. Although an extensive set of investigations is detailed in this document, it is recommended that clinicians use their judgement in individual cases as to which tests should be given priority to ensure optimal diagnostic yield with least intervention.

Guideline Review

The information provided in this guideline will be reviewed by this group and where appropriate updated in line with new evidence and opinion in December 2016.

Funding

The charity WellChild has funded this initiative to develop these recommendations following their funding of the British Paediatric Surveillance Unit Study of the Incidence of Sudden Unexpected Postnatal Collapse.

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Protocol

In the following section recommendations are either referenced or are asterisked (*) where based on expert opinion or consensus of the steering committee.

1. Collapse *outside* hospital

These cases should be investigated as per the guidance for the investigation of Sudden Unexpected Death in Infancy (1) and as per any local procedures. The following additional investigations are recommended in this early neonatal group due to the temporal proximity to birth as well as the contribution of congenital anomalies or infection to aetiology (15). The investigations should also be carried out for survivors.

2. Information to be collected by medical staff following presentation (Appendix 1 for recommended data fields) (*)

This information should be collected as soon as possible after presentation so that all relevant staff are available to give a history and so that memory of events has not deteriorated with time.

1. Parental medical history
2. Full parental drug, alcohol and nicotine history
3. Three generation family tree (noting where available egg donation, sperm donation)
4. Obstetric history including information about infection, fetal growth, suspected fetal anomalies, fetal movements and liquor volume- recommended that this history be sought from a consultant obstetrician or senior trainee
5. Labour and birth, including maternal medication and markers of fetal wellbeing (scalp pHs, cord pHs, CTG, passage of meconium, requirement for resuscitation)- recommended that this history be sought from a consultant obstetrician or senior trainee
6. Health of infant until collapse including growth and feeding
7. Circumstances surrounding collapse, including who was present, whether baby was feeding, position of baby (detailed history obtained from staff who were present at the time of collapse and family
It is also important to collect information from other agencies who may have been involved with the family such as primary care, social care and police
8. Full resuscitation details
9. Full examination

3. Investigations to be performed whilst infant is alive (*)

Liaison with local and regional laboratories is mandatory to ensure optimal collection and timing of samples. Clinicians should use their judgement in individual cases as to which tests should be given priority to ensure optimal diagnostic yield with least intervention (Appendix 4). Transfer to a specialist unit for imaging should be considered if the baby is sufficiently stable.

1. Placenta- where available, both fixed and fresh samples of placenta and cord should be sent as soon as possible after birth for pathology and microbiology.
2. Maternal blood: Kleihauer, viral titres (serum to be frozen for acute phase titres), toxicology
3. Maternal high and low vaginal swabs
4. Neonatal blood:
 - a. Full blood count, coagulation, blood gas, renal and liver biochemistry, glucose, lactate, calcium, magnesium, ammonia, beta-hydroxybutyrate, amino acids, insulin, free fatty acids, acyl carnitine profile, urate, uric acid, cortisol (3 samples at different time points), culture, viral titres, blood spot for cardiolipin analysis
 - b. Specific genetics- DNA and chromosomes, retained blood spot.

- If there is any suspicion that the collapse or death may have been as a consequence of unrecognised hypoventilation / apnoea, then a sample of DNA should be sent specifically to look for abnormalities of the PHOX2B gene which is commonly implicated in congenital central hypoventilation syndrome.
 - Testing for mutations and copy number variation in MECP2 should be considered as this may present as newborn encephalopathy and/or apnoeas and respiratory collapse.
 - Array-based comparative genomic hybridisation is a useful investigation that will replace conventional karyotyping in the near future as a method for detecting causative chromosomal deletions and duplications.
5. Cerebrospinal fluid: biochemistry, glucose (paired with plasma glucose), culture, virology, lactate, amino acids including glycine, storage
 6. Surface swabs: bacteriology
 7. Nasopharyngeal aspirate: bacteriology and virology
 8. Urine: bacteriology, virology, toxicology, organic acids including orotic acid, amino acids including urinary sulphocysteine and urine to be retained for storage
 9. Imaging: skeletal survey (Appendix 2) cranial ultrasound scan day 1, MRI brain (initial and in second week, see Appendix 3 for recommended sequences), renal/adrenal USS, electrocardiogram (at presentation and after 3 weeks of age), echocardiogram
 10. Ophthalmoscopy/ Retcam
 11. Skin biopsy for fibroblast culture
 12. Muscle biopsy if unable to exclude neuromuscular or mitochondrial disorder
 13. Electroencephalogram
 14. Genetics assessment and clinical photographs

4. Investigations performed after death

Where death has occurred *after discharge from hospital*:

These cases should be investigated as per the guidance for the investigation of Sudden Unexpected Death in Infancy (1) and as per any local procedures. The following additional investigations are recommended in this early neonatal group due to the temporal proximity to birth as well as the contribution of congenital anomalies or infection to aetiology.

Where death has occurred on hospital premises:

a) Before post mortem

It is recommended that if it has not been possible to take samples during life then, where feasible, certain samples should be taken immediately following death whilst awaiting post mortem (1). This would prevent significant degradation of material which will occur after death such that important diagnostic information will be lost.

The taking of post mortem samples must be performed on licensed premises (Human Tissue Act 2004) requiring the infant to be taken to the pathology department or where the local Pathology Licence permits, on the neonatal unit or in the emergency department. Each area must establish a local agreement with their Coroner/PF regarding the taking of such samples. Consent should be sought from parents (or Coroner/PF if the case has been referred to Coroner/PF) and documented using the appropriate sections of the standard neonatal post mortem consent form following full explanation of what samples are required and why there is a need.

The baseline samples should, where possible (and always where death occurs in Scotland), be discussed with and agreed by a pathologist and where indicated a biochemist. Those usually recommended are (1):

- Throat and nose swabs for bacterial and viral culture
- Blood culture
- Blood and urine for metabolic studies including glucose, acyl carnitine, organic and amino acids including orotic acid and sulphocysteine, freeze urine for storage
- Blood for DNA, chromosomes and dried blood spots on several cards
- CSF obtained by lumbar puncture or ventricular tap- biochemistry, glucose, culture, virology, lactate, amino acids including glycine, freeze and storage
- Skin biopsy for culture and storage of fibroblasts- 3x 2mm full thickness collected under sterile conditions into culture or viral transport medium or saline soaked gauze. Send promptly to cytogenetics laboratory.
- Muscle biopsy for electron microscopy, histopathology and enzymology- wrap in aluminium foil, snap freeze and store at -70C. Contact metabolic physician or pathologist before collection of sample.

b) Post mortem procedure

Every unexpected death must be reported by law to the Coroner/Procurator Fiscal.

Every death resulting from an unexpected collapse where the cause is not known must be notified by law to the Coroner/Procurator Fiscal. In this situation, the doctor caring for the infant must not issue a certificate of the cause of death, even where that death may be secondary to the hypoxic-ischaemic consequences of the collapse.

It is important to recognise the additional distress that referral to a Coroner/Procurator Fiscal may cause parents and to communicate this sensitively with them, emphasising the routine nature of this process with an explanation as to why the referral is being made (18). It is also important that parents do not feel they are under suspicion for their child's death and instead answers are being sought which may influence future decision-making. Most parents will want to know as much as possible about why their baby died but some find the idea of a post mortem difficult to contemplate even when they acknowledge the potential benefits (19).

- Where the Coroner/Procurator Fiscal orders a post mortem the Coroner/Procurator Fiscal should be urged to have a perinatal or paediatric pathologist perform the examination (1)(16).
- Where there is clinical suspicion of the cause of collapse and the Coroner/Procurator Fiscal does *not* order a post mortem, it remains crucial that the clinical cause of death is confirmed by a post mortem examination. It is highly recommended that such consent be obtained by a consultant paediatrician (*).

It is highly recommended that the post mortem is carried out by a perinatal or paediatric pathologist and that brain pathology is conducted and interpreted by a neuropathologist with appropriate perinatal expertise (1). The post mortem examination should be carried out within 48 hours of the infant's death whenever possible (*). It is essential that all relevant information is available to the pathologist at the time of post mortem to inform the examination. This will include details of the mother, her pregnancy and labour as well as those of the infant, his birth, the events surrounding his collapse and his care until death. It is recommended that pathologists undertaking such examinations liaise with their local

radiology and laboratory colleagues with respect to the range of ancillary investigations as there may be some variation in local practice (*).

Whole organs, other than the brain, will not be routinely retained unless recommended by the pathologist. However, consent should be sought for retention of the brain, and for any other samples or organs which may be useful in refining the diagnosis or in furthering present or future research into the causes of sudden death in neonates (*). If consent for whole brain retention is withheld, the brain may be fixed for 1-3 days before comprehensive sampling of key brain areas, as given below, with retention also of frozen samples as below. Residual tissue, constituting the bulk of the brain, can then be replaced in the infant's body before release for the funeral. It is recommended that as in standard post mortem procedures, consent be sought for use of residual material in tissue blocks and frozen samples for potential future research purposes. Although requesting such consent may be regarded as too sensitive an issue at the time of death, various groups have achieved parental consent rates approaching 100% in such situations (20)(21).

Specific consent will also be required for genetic testing. In the event of non-consent or lack of suitable genetic material from the infant it is important to inform parents that it might be possible to offer them limited genetic investigations. This includes carrier testing for common mutations for some inborn errors of metabolism. Specific advice should be sought from the clinical geneticist.

If despite all efforts both the Coroner/Procurator Fiscal and parents decline post mortem pathological examination, consideration should be given to requesting a post mortem MRI (*).

Procedure

1. Photographs- full frontal, face, profile, any dysmorphic features
2. Radiology- full radiological skeletal survey (Appendix 2), reported by a radiologist with paediatric training and experience
3. Anthropometric measurements- body weight, crown-rump, crown-heel, heel-toe and occipitofrontal circumference
4. Microbiology- to be taken as early as possible during the autopsy procedure and with stringent efforts to avoid contamination.
 - a. Bacteriology
 - i. Blood culture
 - ii. Lung tissue/fluid
 - iii. Bronchial swab
 - iv. Cerebrospinal fluid
 - v. Spleen
 - vi. Any apparent site of infection
 - b. Virology (PCR and culture)
 - i. Postnasal swabs
 - ii. Cerebrospinal fluid
 - iii. Lung tissue
 - iv. Heart muscle
 - v. Small intestine
 - vi. Blood
5. Organ systems- full dissection with weights
Minimum samples to be taken:
Thymus
Thyroid
Larynx

Trachea
Lung (two blocks from each of the lung lobes)
Heart (paraffin blocks to include parts of both ventricles, a major coronary artery and conducting system: sinoatrial node at the superior cavo-atrial junction, the AV node, the bundle of His and bundle branches)
Distal oesophagus
Diaphragm
Stomach
Small intestine
Colon
Mesenteric and subcarinal lymph nodes
Liver (right and left lobe)
Pancreas
Spleen
Right and left adrenal glands
Right and left kidneys
Gonad
Left fifth rib (to include costochondral junction and attached intercostals muscles)

In addition any lesion, injury or congenital abnormality should be sampled independently.

6. Frozen sections- staining with Oil Red O for fat

Liver
Heart
Kidney
Right and left lung
Skeletal muscle

7. Biochemistry

Blood- glucose, urea, sodium, toxicology for maternal illicit drug use, blood spot for acyl carnitine profile and cardioplipin analysis
CSF- sodium, urea, glucose
Vitreous fluid- sodium, urea, glucose, osmolality
Urine- toxicology, amino and organic acids
Bile- bile spot for acyl carnitine

8. Molecular/ cytogenetic investigations

Skin- culture medium
Spleen- frozen
Thymus- frozen
Liver- frozen

9. Retention of material for recognised and unrecognised future purposes

10. Central nervous system examination

It is recommended that a post mortem MRI of the brain be performed prior to opening the cranium (Appendix 2)(22).

Detailed examination of the central nervous system is crucial and requires optimal fixation of the brain as well as retention of frozen samples (see (d) below). The principles of neuropathological

examination require a period of fixation of up to three weeks for optimal studies. Shorter fixation periods may be used where early release of body and its organs for funeral is required.

- a) The fixed brain should be sliced in the coronal plane at 1cm intervals.
- b) All brain slices should be photographed to provide a permanent record of macroscopic appearances.
- c) Block selection
 - Cerebrum:
 - Representative sections of frontal, parietal and occipital lobes.
 - Hippocampus including inferior temporal lobe (bilateral)
 - Thalamus (bilateral)
 - Basal ganglia at the level of the mamillary bodies (bilateral). Many of these blocks can be obtained from a coronal section of the cerebral hemisphere taken through the mamillary bodies
 - Cerebellum:
 - Each hemisphere including dentate nucleus
 - Vermis
 - Brainstem:
 - Midbrain, pons and medulla (all levels of the brainstem if possible)
 - Spinal cord:
 - Cervical, thoracic, lumbar and sacral cord if an abnormality is suspected.
- d) Frozen samples of brain: 1cm³ sample of frontal and temporal lobes, cerebellum and brain stem.

5. Reporting

Reports should be made available as quickly as possible without compromising quality. A provisional report recording macroscopic appearances and all investigations initiated should be made within one week of autopsy to the Coroner/Procurator Fiscal who should thereafter report findings to the clinician (16). Such information is unlikely to be complete and caution is advised if clinicians decide to release such information to parents at this early stage. From the date of autopsy to issuing a final report should normally take no longer than two months.

The lead paediatrician must meet with the parents at the earliest opportunity to explain the findings of all investigations (16). Where the Coroner/Procurator Fiscal is involved, this meeting should be with their consent.

The approach to reporting such infants will be governed by national reporting classification systems but the following is proposed (*)

1. if there is a definite diagnosis then this should be given as the cause of death on the death certificate
2. in all other cases a 'holding' diagnosis should be given eg 'unexplained pending further examination'
3. a final cause of death i.e. a specific diagnosis or 'sudden unexpected neonatal death' should be submitted after all investigations are completed.

6. Final case review meeting

As for all unexpected deaths in childhood the final event in the investigative process should be a detailed local case review meeting, involving all professionals who have been involved in the care of the infant and mother (23). In addition to the pathologist and any relevant professionals from other

agencies, it is essential that this review include a clinical health professional in addition to the pathologist (16). The purpose of this meeting is to share all available information and to reach a consensus on the “cause” of death if possible as well as identifying any contributory or notable factors that did not directly affect the processes leading to death but may be of clinical, social or genetic relevance. As for all unexpected deaths in childhood, understanding and attributing significance to such factors and reaching a conclusion on the likely cause of death or factors contributing to the death requires effective multiprofessional discussion and information sharing. The outcome of such a discussion meeting will be of great value both to those informing the family and to the conduct of a subsequent Crown investigation if held.

7. Communication

Sudden unexpected collapse of their previously well baby is devastating for parents. It is common for parents to feel guilty that they did not recognise warning signs at an earlier stage or that they were in some way responsible for their child’s deterioration. This is especially true where babies collapse whilst breast-feeding or whilst experiencing skin-to-skin with their mother.

Parents should be given appropriate support whilst their baby is undergoing stabilisation and further management. A high quality of communication is required to counsel parents about the current status of the child and to allow sensitive questioning around the situational circumstances of the collapse (24). Parents who are shocked, distressed and frightened are likely to be particularly sensitive to the way staff communicate with them (18).

Parents should be made aware that such collapse is rare but recognised. There are a variety of potential underlying causes and many different investigations may be required to establish aetiology. These will not be performed indiscriminately but will be guided according to the history, examination and progress. Parents should also be aware that even with thorough investigation a cause for collapse might not be found. In such cases parents should have the opportunity to consent to retention of samples for potential future diagnostic or research purposes.

Standards for communicating with parents

- Consistent clear and sensitive information should be provided regularly to parents about their child’s medical status, investigatory process and prognosis where known. In non-emergency situations parental involvement in decision-making is expected.
- Parental contact with their baby should be encouraged and intention to breast-feed recognised and supported where appropriate
- Privacy in suitable surroundings where parents cannot be overheard or seen is important for all sensitive discussions with parents. This is especially important when breaking bad news.
- An awareness that shocked and distressed parents will find it challenging to absorb new and difficult information and may need time to absorb information, and to formulate and ask questions. They may also need to have information repeated.
- Written information should be available, wherever possible.
- The justification for investigations should be explained clearly to parents and consent taken where possible particularly where there is a requirement to transfer the infant to another hospital eg for imaging, electrophysiology
- Results of all investigations should be communicated to parents as soon as these become available

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Appendix 1

Datasheet for investigation of newborn infants who suffer a sudden and unexpected postnatal collapse in the first week of life

Name of person completing form

Grade.....

Contact details.....

Date form completed

History obtained from

1.....

2.....

3.

A. Parental details

Ensure that history from mother or from maternity records matches that held by primary care

1. Maternal details

Name

Date of birth

Hospital number

Current marital status: Married/single unsupported/ single supported (circle)

Occupation

Country of birth

Ethnic group

Pre-pregnancy conditions or illnesses including infertility

Number of previous miscarriages.....

Number of previous terminations.....

Number of previous stillbirths

Number of previous live births

Any previous infant deaths?

Any relevant history in siblings including current health status

.....

Do the family have contact with social services?

Tobacco

How many cigarettes a day before pregnancy

How many cigarettes a day during pregnancy

Was the mother a smoker at the date of birth?

Was there any recent tobacco use in the twenty-four hours prior to collapse?

Alcohol

How many units a week during pregnancy?

How many units in the 24 hours before the baby's collapse?

Illegal substances

Which drugs?

How many times during pregnancy?

What drugs were taken in the twenty-four hours prior to collapse?

If yes, which drug, and when taken in relation to collapse

2. Paternal details

It is important to enquire sensitively whether the putative father is the biological father

Father's name.....

Father's date of birth.....

Occupation

Country of birth

Ethnic group

Tobacco

How many cigarettes a day ?

Was there any recent tobacco use in the four hours prior to collapse?

Alcohol

How many units a week?

How many units in the 24 hours before the baby's collapse?

Illegal substances

Which drugs?

How often?

Was there recent drug abuse in the twenty-four hours prior to collapse?

If yes, which drug, and when taken in relation to collapse

Medical conditions or illnesses

3. Family history of both parents

(eg consanguinity, previous neonatal deaths/ genetic conditions/neurological or thrombotic conditions)

.....
.....

Three generation family tree (inclusive of assisted reproductive techniques):

B. Pregnancy

Last menstrual period

Expected date of birth.....

Gestational age when first booked

Maternal body mass index at booking?

Was this a twin pregnancy at any stage?

If twin pregnancy what was/is the chorionicity?

Was there any concern during pregnancy regarding fetal growth?

a. Was the growth thought to be poor?

b. Was the growth thought to be excessive?

c. Is there information about Doppler, liquor volume, biophysical profile?

.....

Was there any fetal anomaly queried at any stage?

.....

.....

Concern or illnesses during pregnancy (including loss of fetal movement).....

.....

.....

.....

Medications taken during pregnancy (prescribed or over the counter)

.....

.....

Did the mother fall or have any accident during this pregnancy?

.....

Were both parents well at the time of birth and at the time of infant collapse?

.....

C. Labour, delivery and birth

Onset of labour: spontaneous/no labour/ induced If induced, what method?

If not spontaneous please give reason for delivery

Was there spontaneous rupture of the membranes?

How long had membranes ruptured before birth? (hours).....

Length first stage Length second stage.....

Mode of birth: SVD/ breech vaginal/ forceps/ Ventouse/ elective CS/ emergency CS

If not SVD, please give reason for mode of birth.....

List of all medications received during labour including epidural, spinal and general anaesthetics

.....

List of all procedures performed during labour

.....

Were there concerns about the status of the fetus prior to birth?

a) Fetal heart rate

b) Fetal movements

c) Liquor volumes

d) Meconium staining of liquor.....

e) Maternal infection (eg pyrexia, positive swabs).....

f) Vaginal bleeding.....

g) Scalp pH

Lowest fetal scalp pH Base excess Lactate

Cord arterial pH Base excess Lactate

Cord venous pH..... Base excess Lactate

Management of third stage of labour

Placental weight (g) Appearance

Estimated maternal blood loss (ml)

Other notes:.....

.....

.....

D. Infant details

Infant's name

Hospital number

Date and time of birth

Gender.....

Gestation (weeks).....

Birthweight (g)

Head circumference (cm)

Order of birth if multiple

Apgar scores at birthat 1minat 5minat10 min

Time to spontaneous respirations (mins)

Grade of most experienced person at resuscitation at birth.....

What resuscitation was required at birth?

.....
.....
.....

E. Circumstances of collapse

Date of collapse..... Time of day (24 hr clock)

Time baby was last seen or heard to be well (or alive)? (24 hr clock).....

Description of baby at that time

In which ward was baby found collapsed?

What health professionals had been involved in the baby's care at any time prior to collapse?
.....

Who was in the room at the time of collapse?

Who was responsible for supervision of baby just prior to collapse?

Who identified the baby's poor condition?

With whom was baby when found collapsed?

What was the baby wearing at time of collapse?

What was the baby's bedding at time of collapse?.....

Location:	In carer's arms	Position of baby	Prone
	On carer's chest	when identified:	Supine
	On carer's abdomen		On right side
	At breast		On left side
	In mother's bed		
	In own cot	Was the baby	Feeding
	Other	thought to be:	Awake, not feeding
			Asleep

Feeding at time of collapse:

If not feeding 1. baby had not fed since birth

2. breast fedhmins pre-collapse

3. bottle fedhmins pre-collapse

If baby was feeding at time of collapse, was it at breast, by bottle, cup, syringe or nasogastric tube?

.....

What types of milk had the baby received since birth?

.....

Had there been any concerns about feeding prior to collapse?

.....

Was the mother receiving or had she received in the previous 8 hours any medications (including over the counter medications), analgesia (including epidural/spinal anaesthesia) or sedation?

If yes, please provide detail

Was the mother undergoing any procedures at the time of collapse eg episiotomy, epidural top up?

If yes, please provide detail

What was the mother's level of arousal at time of collapse (eg asleep, sedated, exhausted)

Had the baby received any medications/immunisations since birth?

.....

Had there been any concerns about the baby prior to collapse (eg breathing, colour, cry, vigour)?

.....

Had any birth defects been identified prior to collapse?

Had any of the following been a problem prior to collapse?

a) Jaundice

b) Infection

c) Hypoglycaemia

d) Polycythaemia

e) Poor feeding

f) Other.....

Condition when first found collapsed: Respirations breaths/min

Heart rate bpm

Colour

Tone.....

Resuscitation details following collapse (Including time to heart rate > 100, time to first respirations, IPPV/ intubation/ cardiac massage/ resuscitation drugs, injuries observed, oronasal haemorrhage)

.....

.....

.....

.....

Names and grades of all present at resuscitation?

.....

.....

Resuscitation successful? Yes, transferred to NNU at time:.....

No, baby died before or at resuscitation

F. Following collapse

First blood gas after resuscitation: Arterial/ venous/ capillary

Date: Time

pH or H+

pCO₂

BE or bicarbonate

Lactate

First weight of baby and date after collapse Centile

First head circumference and date after collapse..... Centile.....

Length of baby Centile.....

Meconium staining of skin present

Details of full examination (including diagrams where necessary):

G. Investigations to determine the cause for collapse

Maternal:	Date and time	Result
Placenta histology
Blood Kleihauer
Viral titres
Storage
Toxicology
High/low vaginal swabs
Infant:		
Blood FBC
Coagulation
Blood gas
Renal function
Liver function
Glucose
Lactate
Calcium
Magnesium
Ammonia
beta-hydroxybutyrate
Amino acids
Insulin
Free fatty acids
Acyl carnitine profile
Urate
Uric acid
Cortisol (3 diff samples)
Culture
Viral titres
DNA and chromosomes
Dried blood spot (biochem)
Storage (cytogenetics)
CSF Glucose
Protein
Culture
Virology
Lactate
Amino acids
Storage

Surface swabs, bacteriology
Nasopharyngeal aspirate
Urine		
Bacteriology
Virology
Toxicology
Organic acids
Orotic acid
Amino acids
Sulphocysteine
Storage
Imaging		
Chest X-ray
Skeletal survey
Cranial USS day 1
MRI brain (x2)
Renal/adrenal USS
Echocardiogram
Ophthalmoscopy
Other		
ECG (x2)
EEG
Skin biopsy
Muscle biopsy
Genetics assessment
Clinical photographs
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.....
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.....
.....

Post Mortem

.....

.....

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.....

.....

Appendix 2

Standards for skeletal survey in suspected non accidental injury (NAI) in children (25)

A single film ('baby gram') should be avoided as it gives an unsatisfactory exposure and combined views of chest abdomen pelvis and limbs should also be avoided, as limb detail is poor, with oblique projections of most joints.

Skull (SXR)

AP and lateral, plus Towne's view for occipital injury.

SXRs should be taken with a skeletal survey even if a CT scan has been performed.

Body

AP/frontal chest (including clavicles)

Oblique views of the ribs (left and right)

AP Abdomen with pelvis and hips

Spine

Lateral spine - cervical and thoracolumbar

Limbs

AP humeri, AP forearms

AP femurs, AP tibiae/fibulae

PA hands and DP feet

Supplemented by:

- Lateral views of any suspected shaft fracture.
- Lateral coned views of the elbows/wrists/knees/ankles may demonstrate metaphyseal injuries in greater detail than AP views of the limbs alone. The consultant radiologist should decide this, at the time of checking the films with the radiographers

Appendix 3

Protocol for MRI examination of brain

Essential

- T1 and T2 weighted axial images, slice thickness 4mm (use “neonatal” angle- from inferior frontal lobe to torcula)
- T1 weighted sagittal, with thinner slices 1.5-3mm. Volume sequence is ideal as this can then be reformatted into coronal or transverse planes if needed for later comparison with histopathology.
- T2 coronal
- Diffusion weighted imaging with a generated ADC map

Desirable

- MR Sinus venogram
- MR proton spectroscopy in the basal ganglia/thalami; (Echo time 135ms and 270 ms)
- MR Angiography
- FLAIR (optional)
- If strong suspicion of focal infection or herpes encephalitis and normal renal function consider using a low risk contrast agent

Post mortem protocol

Use T1 and T2 weighted sequences from essential list above. T2 weighted images will have best quality. Do each sequence in all three planes. Increase doubling signal averages if time allows to provide a better signal to noise ratio.

Appendix 4

List of conditions described in Sudden Unexpected Postnatal Collapse or collapse in infancy and relevant investigations

Condition	Range of investigations to detect conditions in each category
Infection Systemic: Bacterial infection- various Viral infection- Echovirus, Coxsackie, Respiratory Syncytial Virus, Parvovirus, Herpes Meningitis: bacterial, viral	Placenta-histopathology, bacteriology Maternal blood-viral titres Maternal high and low vaginal swabs- bacteriology Blood- culture, viral titres, storage CSF- bacteriology, virology, biochemistry, glucose Urine- bacteriology, virology Surface swabs- bacteriology Nasopharyngeal aspirate- bacteriology, virology
Cardiac anomalies Cyanotic heart disease - Transposition of the great arteries - Truncus arteriosus - Univentricular heart - Pulmonary stenosis/atresia - Tricuspid atresia Left sided obstructive lesions: - Coarctation/interruption of the aorta - Hypoplastic left heart - Aortic stenosis Cardiac conduction problems - Long QT syndrome - Atrial fibrillation Total anomalous pulmonary venous drainage Myocardial infarction Cardiomyopathies Barth syndrome Congenital coronary artery aneurysm Anomalous coronary artery	ECG Chest X-ray Echocardiogram Blood- Chromosomes and /or aCGH, DNA, storage Genetics for cardiac conduction disorders/cardiomyopathy Skeletal survey Blood spot for cardiolipin analysis
Respiratory Conditions Pneumonia Pulmonary hypertension Pulmonary haemorrhage Aspiration Accidental smothering Congenital diaphragmatic hernia Maternal opiate-related	See investigations for infection Chest X-ray ECHO Endotracheal secretions- bacteriology
Haematological Anaemia	Full blood count and film Maternal Kleihauer Infant and maternal viral titres Placental pathology
Endocrine Hypoglycaemia Hyperinsulinism	Blood- glucose, lactate, ammonia, beta-hydroxybutyrate, amino acids, insulin, free fatty acids, acyl carnitine profile, urate, uric acid

Endocrine (continued) Congenital adrenal hypoplasia	Cortisol (3 samples at different time points), electrolytes Renal and adrenal ultrasound scan
Metabolic Hypoglycaemia Hypocalcaemia Hypomagnesaemia Fatty acid oxidation defects- including MCAD deficiency, VLCAD deficiency, LCHAD deficiency, carnitine acylcarnitine translocase deficiency, carnitine palmitoyltransferase 2 deficiency, trifunctional protein deficiency Urea cycle defects Organic acidurias Lysosomal storage disorders- I-cell disease Peroxisomal disorders- Zellweger syndrome Glycogen storage disorder types 2 or 4 Heart-specific phosphorylase kinase deficiency Mitochondrial disorders- respiratory chain, Leigh's disease Congenital defects of glycosylation Congenital lactic acidoses Glycine encephalopathy Biotinidase deficiency Glucose transporter defect- GLUT1 Molybdenum cofactor deficiency Sulphite oxidase deficiency	Blood- glucose, gas, lactate, ammonia, beta-hydroxybutyrate, amino acids, insulin, free fatty acids, acyl carnitine profile, urate, uric acid, cortisol (3 samples at different time points), VLCFAs, calcium, magnesium, renal and liver biochemistry, DNA and chromosomes, blood spot Cerebrospinal fluid- lactate, amino acids including glycine, storage Urine- organic acids including orotic acid, amino acids including urinary sulphocysteine and urine to be retained for storage Skeletal survey Muscle biopsy Skin biopsy- fibroblast culture Ophthalmoscopy/ Retcam MRI brain Electroencephalogram ECG Echocardiogram
Neurological Any metabolic cause of seizures/apnoea Drug withdrawal Perinatal infarction Intracranial bleed Antenatal injury Hypoplasia of brainstem nuclei Hyperekplexia: apnoea/tonic Congenital hypoventilation syndrome Rett syndrome variants Joubert syndrome	As above (metabolic) EEG/aEEG/cerebral function monitor Maternal and infant urine and blood toxicology Coagulation screen Cranial ultrasound scan MRI brain Blood, skin biopsy: - PHOX2B sequencing (congenital hypoventilation syndrome) - MECP2 sequencing and copy estimation (Rett syndrome variants)
Neuromuscular/skeletal Non accidental injury Congenital myasthenia syndromes Nemaline myopathy X-linked myotubular (centronuclear) myopathy Central core disease	Cranial ultrasound scan MRI brain Skeletal survey Genetics for congenital myasthenic syndromes DNA and chromosomes and/or aCGH Muscle biopsy - single genes screens for MTM1 (XLMM), RYR1 (central core disease), NEM1 -5 (nemaline myopathy), CHRNE & CHRN1 (subunits of the acetylcholine receptor) Ophthalmoscopy/ Retcam (EMG, nerve biopsy)

Appendix 5

Membership of the Steering Committee

Dr Julie-Clare Becher	Consultant Neonatologist, Simpson Centre for Reproductive Health, Edinburgh (Chair)
Dr Michael Ashworth	Consultant Paediatric Pathologist, Great Ormond St Hospital, London
Professor Jeanne Bell	Department of Neuropathology, University of Edinburgh
Miss Charlotte Bevan	Lay representative, Sands, the Stillbirth and neonatal death charity
Professor R Neil Dalton	WellChild Laboratory, King's College London, Evelina Children's Hospital, London
Dr David Fitzpatrick	Consultant in Clinical Genetics, MRC Human Genetics Unit, Edinburgh
Professor Peter Fleming	FSID Research Unit, Institute of Child Life and Health, St Michael's Hospital, Bristol. Member of the Kennedy committee and the working group 'Working together to Safeguard Children'
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Dr Amaka Offiah	Clinical Senior Lecturer, University of Sheffield and Honorary Consultant Paediatric Radiologist, Sheffield Children's Hospital
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Conflicts of interest sought and none declared

Appendix 6

Organisations and representatives involved in the consultation process

Coroners' Society	Dr Roy Palmer, HM Coroner Southern District of London
Crown Office and Procurator Fiscal Service	Ms Linda Cockburn
Scottish Cot Death Trust	Dr John McClure, Chairman Mrs Fiona Brown, Executive Director
Bliss*	Mr Andy Cole, Chief Executive
International Stillbirth Alliance*	Mr Ron Gray
National Childbirth Trust*	Ms Margaret Mannion
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Royal College of Pathologists*	Dr Michael Ashworth
Royal College of Paediatrics and Child Health*	Professor Terence Stephenson Clinical Standards Committee
Scottish Pathologists Association*	Dr Jean Keeling
British Association of Perinatal Medicine*	Dr Bryan Gill
British Neuropathology Society*	Professor Jeanne Bell

* Organisations involved in external review of the guideline

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