

Technical method

Pathological investigations in cases of sudden infant death

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There are currently two extremes of approach to the pathology of sudden infant death syndrome (SIDS). One is represented by the traditional approach of the coroner's pathologist which aims to exclude any "unnatural" form of death by macroscopic dissection, but entails little or no microbiological or histological study. In contrast, is the approach used in research programmes such as the multicentre post-perinatal mortality study.¹ This includes whole body radiology, multiple sampling for bacterial and viral cultures, blood studies for immunoglobulins, and viral antibodies and aspiration of the vitreous humour for biochemical examination, in addition to extensive dissection and multiple sampling for histology of most internal organs. The latter approach is completely open ended as there is no limit to the range of sites that can be sampled for any technique, or to the steady stream of new hypotheses which suggest further biochemical or microbiological hares to be chased. Those paediatric pathologists whose main research is on SIDS will not find this a problem. For pathologists who work in district general hospitals and academic pathologists who undertake research into areas other than SIDS, the problem is very real.

Despite the published reports on the amount of underlying pathology that may be shown by detailed necropsy in SIDS,¹⁻³ the proportion of cases where extended studies yield information of practical value to the family or their medical advisers is relatively low. This is in direct contrast to what happens in fetal and neonatal pathology where detailed studies yield information which explains the death or leads to a planned approach towards prevention of recurrence in a high proportion of cases.⁴ Moreover, in the in-

vestigation of a perinatal death a diagnostic algorithm can be followed so that no more work is undertaken than that required to achieve the desired purpose.⁵

Faced with the necessity to examine a case of SIDS within a time frame limited by academic or other routine activities, the open ended approach to the SIDS post mortem examination becomes unrealistic even to the paediatric pathologist. He may have to "cut corners" in the examination, delay it until more time is available, or request the coroner to refer the case to another, possibly less well qualified, pathologist. There needs to be some basic standard for investigating cases of SIDS, which can be expanded according to (i) the time or other facilities available, or (ii) the nature of any specific research investigation being conducted.

This protocol was drawn up by a working party of the British Paediatric Pathology Association convened under the auspices of the Foundation for the Study of Infant Death precisely to provide such a basic standard. The authors all regularly conduct post mortem examinations on cases of SIDS at their respective centres.

Necropsy study of cases of SIDS

For an adequate post mortem examination on a case of SIDS the two basic prerequisites are a good clinical history and access to the infant as soon as possible after death.

CLINICAL HISTORY

Although the pathologist usually has to rely on the history provided by the coroner's officers he should attempt to elicit the following details: date and place of birth; gestational age at birth; previous health of the infant and the family; feeding and immunisation history; when the child was last seen by the general practitioner, or other doctor, or health visitor; time the child was last seen alive; circumstances in which the infant was found including clothing and evidence of social condition; details of siblings and whether there have been previous perinatal or infant deaths.

The post mortem examination can be subdivided into two stages.

Stage 1 (initial study and samples to be collected within 12 hours of death)

This can be done in hospital casualty if the infant is brought into hospital (most cases in some areas, very few in others). The studies can be performed when any resuscitation attempts have been abandoned.

They can appropriately be performed by the paediatric staff who should then inform the pathologist of what has been done.

The state of the infant should be noted with details such as the quantity of clothing and presence of froth round nose and mouth. The rectal temperature should be taken and recorded. Samples to be taken if possible (this will not be a problem if resuscitation attempts have been made, but may require cooperation from the coroner if the infant has been certified as "brought in dead") include the following: heart blood (or blood from sagittal sinus if preferred) for bacterial culture; spinal cerebrospinal fluid for bacterial and viral culture; nasal swab or aspirate (check local laboratory preference) for viral studies; if there is evidence (or recent history) of diarrhoea rectal swab for enteropathic *Eschenchia coli* etc.

Stage 2 (standard post mortem study)

Up to 48 hours after death nasal aspirates (if not obtained in stage 1), for viral study and blood cultures for bacteriology are worth while. Cerebrospinal fluid bacteriology is worth doing for at least this period (most will be sterile).

Vitreous humour should be examined for sodium and urea in all cases and, if the post mortem examination is performed less than 12 hours after death, glucose may also be worth measuring.

Internal organ samples should comprise: lung for bacteriology and virology; heart and brain for virology; bowel content for bacteriological and viral studies if there is evidence of diarrhoea.

Fibroblast culture for metabolic studies and storage of fibroblasts for future possible study are justified only if there are features or history suggestive of metabolic disease, or there has been a previous death of a sibling.

Whole body x-ray is useful if the apparatus is available.

NECROPSY DISSECTION PROCEDURES

Measurements to be made and recorded in all cases include: body weight; crown heel length; crown rump length; occipito-frontal circumference. External features should be noted and recorded including state of nutrition, minor anomalies, and evidence of trauma or treatment: particular attention should be paid to the airways.

The internal dissection should follow a standard technique and entail consideration of all systems. The cranial cavity should be opened and the brain removed for examination in all cases, although it is not essential that the spinal cord should be removed in cases with no relevant history or signs of neurological disorder, and examination of the middle ear cavities is a selective procedure.

The major internal organs should all be weighed so that it is subsequently possible to recognise gross deviations from normal. Weights of brain, lungs (left and right), liver, spleen, kidneys (left and right) and thymus should all be recorded, preferably to the nearest 1 g, and adrenals to the nearest 0.1 g. It is immaterial whether the weights are taken in the fresh state or after a brief period of fixation.

The following list of tissues should be sampled for histology as a routine: (i) in the respiratory system the epiglottis, larynx, trachea, main bronchi, lungs—at least four major lobes, as the respiratory system is the most common site of disease and requires extensive sampling; (ii) in the cardiovascular system the left atrium-ventricle, interventricular septum (with related left and right ventricles) should be sampled; (iii) in the alimentary system salivary glands, duodenum with head of pancreas, jejunum, ileum (with Peyer's patch), liver; (iv) and in the reticulo-endothelial system the spleen, thymus, and mesenteric node should be sampled. For (v) endocrines, adrenal and thyroid (with trachea); for (vi) the renal tract, kidneys (one block of each); for (vii) CNS, meninges (with inferior surface of cerebellum), frontal lobe (with centrum semiovale), corpus callosum, head of caudate, and ependyma; and for (viii) the skeletal system, costo-chondral junction (rib 5 or 6), psoas muscle (for longitudinal and transverse sections) should all be sampled.

Defects of lipid metabolism are readily overlooked by the inexperienced. Routine fat stains on samples of liver, kidney, heart and psoas muscle are recommended to exclude the possibility.

It is important that a good quantity of reserve tissue from each organ is kept in fixative until investigations are complete and the post mortem is signed out. The necropsy is not complete until the findings have been phoned to the doctor or other agency caring for the parents.

The above suggested protocol does not cover all eventualities. There is no substitute for an informed necropsy performed by a pathologist who is interested in paediatric problems and to whom minor differences from normal may indicate additional relevant investigations.

The occurrence of a second unexpected infant death in a family, or an unexpected infant death in a family with a previous unexplained neonatal death is an absolute indication for the paediatric pathologist to have a role in the investigation. Additional studies that should seriously be considered in all such cases include toxicological screen, investigation of the cardiac conduction system, and studies of immunological state, in addition to performance of whole body x-ray and preservation of material for metabolic studies.

Problems and additional requirements

During the discussions of the working party several practical problems were identified which may make it difficult for pathologists to follow the protocol in its entirety.

The clinical history obtained by the coroner's officer often omits many of the details suggested above. We think that it would be helpful if the Coroners Society were to provide a standard check list of questions which need to be asked and observations to be made when parents and infants are first seen.

Collection of appropriate samples for microbiological examination within a reasonable time after death requires liaison between the pathologist, the coroner, and the clinicians who first see the infant. On a local basis such a liaison can often be established but in some areas there will be problems outside the control of the pathologist, such as referral of cases of SIDS by general practitioners directly to the public mortuary. Organisations such as The Foundation for the Study of Infant Death and the British Paediatric Association may need to consider this problem further in association with the Coroners Society.

If post mortem examinations are performed at a public mortuary it is seldom possible to weigh organs of paediatric cases as any scales which may be available weigh in increments of 50 or 100 g and are useless for paediatric purposes. The provision of accurate scales for this purpose would be an inexpensive but helpful advance.

There are problems in many areas in the United Kingdom regarding payments for special investigations such as histology and microbiology. It is arguable as to whether these constitute category 2 investigations requiring authorisation of payment by

the coroner, or research investigations requiring payment by a research organisation. We regard it as essential that the need for such studies is agreed and paid for in all cases of sudden infant death.

Finally, although there is an increasing tendency to refer cases of SIDS to pathologists who have an interest in paediatric pathology, this is probably not feasible in all cases. It is important to emphasise, however, as stated above that a paediatric pathologist must be consulted in all second cases of cot deaths or where there has been a previous unexplained neonatal death.

We gratefully acknowledge the encouragement and financial support of the Foundation for the Study of Infant Deaths which enabled the study group to meet and prepare the report on which this publication is based. We are also grateful to Dr Richard George of the Children's Hospital, Birmingham, for checking our suggestions on microbiological investigations.

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