Papers and Originals

A Study in Cot Death*

R. T. COOKE,† M.D., F.C.PATH., R. G. WELCH,‡ M.D., M.R.C.P.

Brit. med. J., 1964, 2, 1549-1554

As the mortality among infants and young children from the well-defined causes falls, so the importance of "cot deaths" increases. When put to bed either these children are well or such symptoms as they have are so trivial that no one can anticipate a fatal outcome; yet a few hours later they are found dead or moribund. In the past (Barrett, 1954) death has been attributed to suffocation by bedclothes, overlaying, or aspiration of vomit. Later it was thought that the child had been overwhelmed by a virulent infection, probably arising in the respiratory tract, and even more recently it has been suggested that death has resulted from an anaphylactic reaction to milk inhaled into the larynx or trachea (Parish *et al.*, 1960). However, the cause of death in most cases remains obscure, and, in the hope that by adding to the evidence already accumulating we might help to clarify the problem, we report here 122 cases of death occurring outside hospital in children between the ages of 2 weeks and 2 years, the circumstances of which were such that the death was reported to the coroner. These deaths occurred in the twin towns of West Hartlepool and Hartlepool (combined population about 90,000) during September 1948 to December 1962 inclusive. All but six of the necropsies were performed by one of us (R. T. C.), and the facts presented are based upon statements obtained by the coroner's officer from the parents, general practitioners, and other persons concerned, together with the gross post-mortem findings and bacteriological and histological studies.

The cases fall easily into two groups. Group A: 31 children with naked-eye evidence of disease or abnormality and, in most, a definite history of illness sufficient, together, to account for death. Group B: 91 children in whom the characteristic findings at necropsy were (1) the presence of petechial haemorrhages in varying number on the surface of lungs and heart and on and in the thymus; (2) the substance of the lungs diffusely congested and slightly indurated; and (3) the trachea and main bronchi typically lined by a fine white froth and similar froth not uncommonly present at the nostrils when the child was first found dead. Besides these abnormalities there may be a patchy pallor of the substance of the liver and kidneys and sometimes opaque mucus or pus in one or both middle-ear cavities. In most of these cases bacteriological and histological investigations have added nothing, but in three children an unexpected abnormality sufficient to account for death was found.

Groups A and B are referred to subsequently as the "definite" and the "typical" respectively, implying either a definite cause of death found or the typical findings as described above without clear cause.

Incidence

Table I shows the incidence by year of the two groups, and relates them to the total number of births, infant deaths, and the number of deaths in the 2-week to 2-year age-group, both as a

whole and as occurring outside hospital. The infant mortality rate, though falling, remains higher than the national average. The deaths in the 2-week to 2-year group have declined steadily, but deaths outside hospital continue at about 12 a year, so that in the past five years 80% of deaths outside hospital in this age-group fall into the sudden and unexpected category. There is some suggestion that the proportion of unexplained deaths is decreasing: of the deaths occurring in the first seven years only 17% were satisfactorily explained, but in the second seven years 33%.

TABLE I.—Incidence of Cot Deaths Since 1950

Year	Total Infant Births	Total Infant Deaths	Total Deaths 2 weeks to 2 Years	Total Deaths in Home 2 Weeks to 2 Years	Total Cot Deaths	Cot Death, Definite Cause Found
1950	1,751	78	50	25	3	1
1951	1,810	71	45	27	11	1
1952	1,802	74	42	19	9	1
1953	1,737	68	33	—	8	1
1954	1,665	75	36	22	12	3
1955	1,811	82	35	22	6	0
1956	1,843	76	31	12	11	5
1957	1,968	86	40	16	7	0
1958	1,963	56	26	11	9	0
1959	1,979	39	21	12	10	5
1960	2,020	64	26	13	8	2
1961	1,975	23	23	11	11	7
1962	2,048	61	27	14	11	3

Two cases were seen in 1948 and four in 1949.

Clinical and Post-mortem Features

Definite Group: Cases With a Convincing Cause of Death; 21 Boys and 10 Girls

Apart from three accidental deaths all save five had a definite clinical illness before death, although 10 had not been under the doctor's care. Five (16%) were under 3 months old, 17 (55%) were over 6 months old. Had they lived a little longer, most of these children would have been admitted to hospital and might have survived. The causes were as follows:

1. *Accidental Death*.—(a) Strangulation by jacket catching on nail (male, 7 months). (b) Concussion by fall from mother's arms on to stone floor (male, 9 months). (c) Nut from chocolate at bifurcation of trachea (male, 18 months).

2. *Gastroenteritis*.—Six males, all under medical treatment and dehydrated at post-mortem examination. *Escherichia coli* 0119 cultured from the bowel of one.

3. *Septicaemia*.—Three meningococcal, one staphylococcal (from abscess of leg). Two died on the way to hospital, one while awaiting the doctor's visit, and the fourth was being treated at home by the doctor.

4. *Acute Meningitis (Haemophilus influenzae)*.—Female, 9 months. Died in doctor's car on the way to hospital.

* C. H. Milburn Prize Essay, 1964.

† Consultant Pathologist, General Hospital, West Hartlepool.

‡ Consultant Paediatrician, General Hospital, West Hartlepool.

5. *Respiratory-tract Illness*.—(a) Acute epiglottitis (*H. influenzae* in heart blood). (b) Bronchopneumonia, three cases (otitis media in one). (c) Purulent bronchitis. (d) Pneumonia with lung abscess (staphylococcal). Four of the six had not been seen recently by a doctor.

6. *Renal Abnormality*.—(a) Unilateral renal-vein thrombosis. Two cases: male, 3 weeks (P.M. blood urea 420 mg./100 ml.); female, 2 months, with otitis media also (P.M. blood urea 660 mg.). (b) Pyelonephritis, meningocele, and lipoma (female, 11 months: P.M. blood urea 370 mg.). (c) Congenital bilateral hydronephrosis and hydroureters and failure to thrive (male, 2 months). Only the third case was having medical attention.

7. *Congenital Heart Disease*.—(a) Fibroelastosis. Two cases: males, 6 weeks and 13 months, dying after a brief illness. The elder had two types of salmonella grown from the bowel. (b) Ventricular septal defect and heart failure (male, 8 months). (c) Ventricular septal defect, mongol (female, 3 months). Only the last child had been seen recently by a doctor.

8. *Miscellaneous*.—A girl of 6 months had not been seen by her doctor since birth, despite prolonged difficulty in feeding; she was thin and dehydrated, and the skin showed numerous chafes and scabbed areas; both middle ears contained frank yellow pus from which pneumococci and *H. influenzae* were cultured; death was attributed to acute bilateral otitis media and malnutrition, and the microscope later also showed acute bronchitis. A girl of 5 weeks was also frail, wizened, and dehydrated, and death was attributed to marasmus and acute congestion of the lungs; her twin brother had been found dead in the twin cot three weeks before with no preceding illness, and an inquest was held without a post-mortem examination being performed. A 9-week-old boy became jaundiced and died suddenly six days later after coughing during a feed—he had neonatal hepatitis.

Typical Group : Cases With Typical Cot Death Findings— 45 Boys and 46 Girls

In this group 60% were under 3 months and 12% over 6 months. In 44 children (24 girls) there was no history suggesting illness whatsoever, and in 10 of these the face was reported as being covered or buried in bedclothes or a pillow when the child was found dead. In the remainder the symptoms reported varied from the trivial to the clinically significant; among the former we include the child who was heard to cough twice and who sneezed a few times in the night, another who had always done a lot of sneezing, and a third who appeared to have recovered from a recent infection of the upper respiratory tract. One child had had pertussis and was still having an occasional bout of coughing at night; another had seemed to be sweating when she took her last feed, and two others woke and cried in the night, but were settled easily; one of these had also vomited. Two more were lifelong vomiters (regurgitators), and in one of these this habit had been more troublesome in the 48 hours before death; the other woke and cried in the night and seemed to be sweating, though he settled after a feed; both these infants had milk in the trachea and bronchi at necropsy. Two infants had cried more than usual in the 24 hours before death, and one of these had been examined by the general practitioner only a few hours before death, the only abnormality found being a tight foreskin. In 14 children a recent cold and/or cough was apparent, but apart from slight restlessness no other sign of distress had been noted by the parents. Another child had been unwell two days before with crying, sweating, and green stools; next day he was apparently normal. *Shigella sonnei* was cultured from his colon after death. All of these children with trivial symptoms were found dead, and in five the face was reported as being covered or turned downwards, and in two more it was doubtful whether the face was completely clear.

The last 21 children had a very definite and unequivocal history of illness preceding death, and 11 were seen to die—four in their cots, two in prams, four while being nursed, and one in the ambulance on the way to hospital. Four died after a fit—following a period of crying and restlessness in one, fever in two, and listlessness, anorexia, and slight diarrhoea in the

fourth. A fifth child found dead had, according to the parents, been having repeated convulsions in the preceding week, although not seen by a doctor. In seven more the history suggested respiratory-tract illness with a varying picture of cough, colds, difficulty or heaviness in breathing, restlessness, lack of appetite, retching or vomiting; only two of these were seen dying—one lapsing into unconsciousness, the other turning pale and dying suddenly. The nine others followed no special pattern save for being unwell for 12 to 24 hours, restless, irritable, refusing to feed, some vomiting, and almost all being described as pale or cold, with sweating. Five of them were seen dying—in two the eyes were described as being open and staring, one was yawning and retching just before death, one breathing heavily, and another gasping. Two of the nine had vomit in the larger air passages at necropsy. It was in this group of children with a definite clinical illness preceding death that the microscope showed unexpected changes contributing to a decision on the cause of death—namely, pyelonephritis in two, bronchopneumonia in one. Two others with a less definite illness also showed microscopical evidence of acute inflammation in the kidneys. In two cases with no clinical illness a few pus cells were found in the lumen of bronchi.

In summary, therefore, with half of these children there was no history of preceding illness, and death came as a complete surprise; in a further quarter there was nothing to suggest that the symptoms noted were important. In 15 the face was reported as being turned down or covered, and in two more there was considerable doubt whether it was clear.

Social and Economic Background

Age at Death and Sex Incidence.—Table II shows that 60% of the children in the typical group were less than 14 weeks old and only 12% were over the age of 6 months, in direct contrast to the definite group where the percentages are 16 and 55 respectively. There is no important sex difference.

Season of Year at Death.—Table III shows that in both groups twice as many died in the six months November to April as in the other six months.

TABLE II.—Age at Death and Sex Incidence

Group	Sex	< 4 Weeks	4–13 Weeks	14–26 Weeks	27 Weeks to 1 Year	1 to 2 Years
Definite	M	1	3	6	7	4
	F	0	1	3	4	2
Total		1	4	9	11	6
Typical	M	3	25	10	4	3
	F	2	25	15	4	0
Total		5	50	25	8	3

TABLE III.—Monthly Incidence of Cot Deaths

Group	July	Aug.	Sep.	Oct.	Nov.	Dec.	Jan.	Feb.	Mar.	Apr.	May	June
Typical	6	3	5	5	5	16	15	7	9	9	5	6
Definite	1	2	1	6	3	2	8	4	1	3	1	1
	6	5	6	11	8	18	23	11	10	12	6	6

Occupation of Father.—This was known in all but five instances, one of which (a Serviceman) is not classifiable. The rest are expressed as percentages, and for comparison the Registrar-General's 1951 census figures for the two towns are shown, also as percentages (Table IV). A distinct preponderance of deaths occurred in families in the lower social grades.

Size of Family.—With the exception of some of the twins, all dead children were the youngest in the family. The size of the family was known in 117 cases (Table V). About a third of the children, therefore, were from families with four or more children, 34.5% of the definite and 33% of the typical group.

Birth Size and Prematurity.—These were not recorded in some of the early cases in the series, but there is nothing in the records to suggest that the delivery was not at term or that the size was unusual. Weight at birth averaged 6 lb. 15 oz. (3,145 g.) for the 27 definite cases where it was known, and 6 lb. 12 oz. (3,060 g.) for the 78 typical cases. In the definite group there were four children of low birth weight—a twin weighing 4 lb. 14 oz. (2,210 g.) at or near term, another twin 4 lb. 3 oz. (1,900 g.) at 36 weeks' gestation, and two singletons weighing 2 lb. 6 oz. (1,075 g.) at 28 weeks and 3 lb. 4 oz. (1,475 g.) at 32 weeks (dying at four and two months respectively). In the typical series there were 15 children of low birth weight, the smallest a twin who weighed 3 lb. 14 oz. (1,755 g.) at term,

TABLE IV.—*Social Grade of Father*

	Social Grade					Not Known
	I	II	III	IV	V	
Definite	0	0	27.5	20.7	51.8	2 cases
Typical	0	4.6	39	16.1	40.3	3 cases
Series total	0	3.4	36.3	17.3	43.0	
1951 Census	2	10	50	14.5	23.5	

TABLE V.—*Position of Child in Family*

	1st	2nd	3rd	4th	5th	6th	7th	8th	9th
Definite	7	6	6	3	—	5	—	1	1
Typical	18	25	16	14	6	5	3	—	1
Total	25	31	22	17	6	10	3	1	2

the next a twin who weighed 4 lb. (1,815 g.) at birth at 37 weeks' maturity. Four more were members of a twin pair, two born at term. Of the remainder at least one other was born at term, the rest with variable degrees of immaturity and weights between 4 lb. 5 oz. and 5 lb. 7 oz. (1,955 g. and 2,465 g.). In contrast with the 19 "premature" infants (15.5%) the prematurity rate for the two towns over a recent five-year period was 8.4%.

Illegitimacy.—Eleven children (including the pair of dead twins) were illegitimate. A twelfth child was illegitimate but had been adopted some weeks before death. Three of the 11 were in the definite series, and the infants and mothers lived with the grandparents. Of the eight in the typical group, four lived under similar circumstances, but the parents of the other four were living together as a family unit.

Associated Abnormality.—Two infants, one from each group, were mongol children.

Previous "Cot Death."—Three families had lost other children under these circumstances. In one, a marasmic infant (placed in the definite series because of her gross wasting) had lost her twin brother three weeks earlier, found dead in the cot, death being ascribed at the inquest to asphyxia (no necropsy). Five years later a 2-months-old child of this family was found moribund and at necropsy there was bilateral hydro-nephrosis and hydroureters. The second family lost children in 1956 and 1960, both in the typical series, and the latter was one of twins. The third family lost children in 1958 and 1962, both typical cot deaths.

Twins.—Many of these infants have already been mentioned, but they are, of course, very important also from the point of view of infection. There were five members of a twin pair in the definite group—enteritis (sibling well), marasmus (sibling dead), concussion, epiglottitis (sibling probably well), and foreign body in trachea. In the typical group there were 11 (six boys and five girls), including one pair: six weighed less than 5½ lb. (2,485 g.) at birth, though only two were more than two weeks early by dates; only one was more than 6 months old (20 months) and eight were from families with four or more children. Two were found moribund, one in a cot with her twin, whose chronic "cold" was unaltered. She was cold, pale, sweating, and gasping for breath, and died in about two hours. At necropsy

stomach contents were found in the air passages. The surviving member of the other pair had been chesty for a few days before the twin began to cough, rapidly became more ill and pale, seemed to catch his breath when he breathed, and died awaiting the doctor's call. Histological examination of his lungs showed some excess of polymorphonuclear leucocytes though no frank pneumonic change. Three died alone in their cots or prams. The surviving twin of one had a cold and cough which worsened after the episode. The second child had been in a local authority residential nursery for seven weeks, her sibling being quite well; and the sister of the third child, though well, had been removed into the parents' bed in the night because she was crying. The other six were sharing a cot or pram. The survivor of one pair, by his crying, called the parents to the cot, but it was near feed-time; three other survivors were apparently well, and one pair were found dead together.

Feeding Habits.—In the definite group only one child (the mongol with cardiac abnormality) was breast-fed, and she had just started on weaning diet. Of the rest, rather more than half had been breast-fed for a while (usually a few weeks only) before changing to cow's milk in one or other preparation. In the typical group four infants were completely breast-fed; another had her first and only cow's milk feed the day before death, and a sixth had an occasional cow's milk feed.

Sleeping Habits and Place of Death.—There was no real difference between the two groups. The normal sleeping-site was not recorded in eight instances in each group, but the place of death was recorded in all: 28% of the infants slept in prams, 44% in cots, 21% in the parents' bed, and eight children in other places—one in an armchair, one on a settee, three in drawers, one in a carricote, and two in an adult bed. As one might expect, in the definite series, where most children were clinically ill, the number of deaths in places other than the normal sleeping-site rose because the children were being nursed or were travelling to hospital. In the typical group much the same thing happened with the children found moribund. In addition the percentage of children found dead in the parents' bed was 5% more than the proportion who usually slept there.

Pathologist's Findings

Nutrition at Post-mortem Examination.—This has been assessed by the pathologist's visual impression and by comparison of measurements, recorded in most instances, with the tables prepared by Tanner and Whitehouse (1959). In the definite group nutrition was regarded as above average in 49%, poor in 29% of the males and in 50% of the 10 females. In the typical series nutrition was again considered to be above average in 42% but poor in 25%.

Presence of Blood Spots on Organs.—These were present, usually in considerable number, in all but five of the 91 typical cases; in three of the negatives there was evidence of inhalation of stomach contents—two fresh and one less recent. In the definite series spots were present in 10 of the 31, and in no case were they numerous. The children showing the spots were three with septicaemia, one strangled, one with the intratracheal foreign body, one with bronchopneumonia, one with purulent bronchitis, and the three with (a) ventricular septal defect and effusions, (b) fibroelastosis of the heart, and (c) recent neonatal hepatitis. The last of these three coughed and went blue, dying soon afterwards; the other two had been briefly unwell before dying.

Presence of Soft Parietal (Craniotabes).—This condition was apparent at necropsy in three of the definite group (two males and one female) and 37 of the typical group (23 females and 14 males). It was found in 21 of the 55 children under 14 weeks old, in 12 of the 25 between 14 and 26 weeks, and in 4 of the 8 aged 6 months to 1 year. In one child the rib ends were examined microscopically, but no evidence of rickets could be found.

Histological Findings.—Microscopical examination of various organs was carried out in 110 necropsies, the exceptions being mainly in the definite group, where the cause of death was apparent, and in some of the early cases. Samples of lung, liver, and kidney were normally studied, and in the last six years heart, adrenals, spleen, and other organs as well. In most of the typical children the lungs showed patchy oedema and intra-alveolar haemorrhage with small areas of emphysema and collapse, with desquamation of the bronchial epithelium either as strips or of the explosive type, and the presence of a few cells of mononuclear macrophage-type in the alveoli. In some there was a slight excess of polymorphonuclear leucocytes in the stroma or in bronchial walls, but not enough to substantiate a diagnosis of infection. In only one child did microscopical examination of the lungs allow a change of diagnosis to one of bronchopneumonia, and this child had had a definite clinical illness before death, and, though regarded as a typical case on gross post-mortem findings, showed only a few blood spots and an unusual colour and feel to the lungs. Unsuspected changes in the kidneys were found in five of 70 pairs examined, and were thought to be the cause of death in two. In one of the latter there were dense and widespread acute inflammatory changes leading almost to micro-abscess formation, and also similar changes in the other with, in addition, quite numerous thrombosed veins. Both these children had clearly been ill before they were seen to die and showed very few blood spots on the intrathoracic organs. In a third child there was a patchy, frank, but not marked excess of polymorphonuclear leucocytes between the medullary tubules; in another there were a few collections of these cells in the outer cortex, with elsewhere a delicate scatter of them between the medullary tubules (in one place quite dense); and in the fifth child there were some areas of nephrosclerosis in the outer cortex with small groups of inflammatory cells in relation to some of them and a few small subcapsular haemorrhages. These last three infants were all found dead, with typical cot-death findings and a non-contributory history.

Bacteriological Findings.—Bacteriological cultures were prepared from 85 children. Most necropsies were carried out on the same day as the child was found dead, between 4 and 12 hours of discovery. The organisms cultured from the typical cases and the correlation with the clinical history are shown in Table VI.

TABLE VI

Organisms Grown	No. of Cases	Clinical Illness Before Death		
		Definite	Doubtful	None
<i>H. influenzae</i>	4	—	3	1
<i>Staph. pyogenes</i>	12	1*	4	7
<i>H. Strep.</i> { Group A, C, or G.	2	—	1	1
{ Not groups A, C, or G.	2	—	1	1
<i>Pneumococci</i>	6	1†	2	3
<i>Str. viridans</i>	3	—	1	2
Sonne dysentery	1	—	1	—

* Had had running ears; profuse growth from bronchi.

† Positive heart-blood culture.

Discussion

Elliott (1961) thought that the diagnosis at post-mortem examination in cases of sudden death in infants, "is not so much a function of what the pathological findings are as of the pathologist who finds them." Our series closely resembles those described by others in recent years, but the interpretations of the findings often differ.

We have found an apparently satisfying cause of death in about a quarter of the cases. Coe and Hartman (1960) found a definite cause of death in 44% of 78 children (bronchopneumonia or laryngotracheobronchitis in 26) and questionable inflammation of the respiratory tract in a further 28%. Valdes-Dapena *et al.* (1963) and Valdes-Dapena and Hummeler (1963), despite exhaustive histological, bacteriological, virological, and

toxicological investigations, found an acceptable cause in only 30% of 114 cases, and they comment that even 10 of these were questionable.

Stowens (1957) found unequivocal signs of disease in 18% of 200 necropsies. Banks (1958), reporting the early findings of a Ministry of Health inquiry into this subject in the London and Cambridge areas, gives the histological classification of the two series of cases; in London death was attributed in 73% of cases to tracheobronchitis, possibly of viral origin and with or without the typical explosive epithelial desquamation as described by Bodian and Heslop (1956); conversely, in Cambridge the cause of death was given as uncertain in 76.5% of cases, the other diagnoses in both series being very similar. Adelson and Kinney (1956) found histological evidence of inflammation of varying degrees of severity in the respiratory tract in 106 out of 126 infants, classifying it as severe in 29 and finding abnormality of the other systems alone or with some degree of respiratory-tract change in some 40 others. The difficulty of interpretation applies even to the gross findings at necropsy. Elliott (1961) reported a series of 31 necropsies carried out by five pathologists. Between them, two pathologists found death in 9 out of 16 infants due to inhalation of stomach contents, and four to infection. The other three ascribed death to infection in 10 out of 15, and in none to inhalation of vomit.

Apart from these more controversial diagnoses of respiratory-tract disease there is close similarity in the conditions found, largely congenital heart disease, meningitis, myocarditis, pyelonephritis, enteritis, and septicaemia, with or without adrenal haemorrhage. In our series we draw attention to the surprising incidence of renal abnormality (in nine children) and to the child who died of acute epiglottitis, from which, as in cases reported by Mann (1961) and Jones and Camps (1957), *H. influenzae* was grown.

There is a remarkable sameness about the infants in our typical group, not only in the gross findings at necropsy and on further microscopical examination, but also in the clinical and socio-economic background. All but 12% were under 6 months of age at death, more than half less than 3 months old. In half there was no history whatsoever suggesting illness; in a quarter what symptoms there were seemed trivial. At necropsy no difference was apparent between the children found with the face covered or with it clear. The hallmark was the presence of blood (Tardieu) spots on the intrathoracic organs; and, indeed, the presence of these in some of the children in the definite group and in conditions where they might not be expected to occur—for example, septicaemia and suffocation by strangulation, impacted foreign body, or extensive infection—suggests that the cause of death at the particular time it occurred might be due to the same mechanism as in the typical series, the abnormalities found being contributory or even coincidental. The presence or absence of milk in the large air passages did not appear to be related regularly to any change in the usual findings.

There is no doubt that aspiration of milk or other food deep into the pulmonary tree can kill, but its presence in the larger tubes is thought more often to be an agonal phenomenon—perhaps delivering the *coup de grâce* in a convulsing or otherwise unconscious child. However, it has been shown (in adults) by Gardner (1958) that gastric contents may gravitate into the lungs and even set up a vital cellular reaction after death. Attempts at resuscitation probably increase this tendency.

Another finding which has interested us has been the presence of craniotabes, a palpable softening of the parietal bones, present in a third of the children. In young infants this is now thought to be a normal variant, but has also been held to be a manifestation of rickets. Recent clinical experience of a very near fatality in a plump twin child of low birth weight, thriving on normal feeds with vitamin supplement, who developed typical laryngismus stridulus at the age of 7 weeks with hypocalcaemia, and who responded to increased dosage of vitamin D, has made us wary of discarding this older belief (Smith and Cooke, 1948).

It can be seen that the histological and bacteriological investigations carried out have been for the most part unrewarding, a satisfying histological diagnosis being apparent in only 3 out of 91 in the undiagnosed group, each of whom was in some ways not truly typical, although no gross findings of other abnormality were recognized. The significance of potentially pathogenic organisms isolated from necropsy material some hours after death must be doubtful. However, it seems not unreasonable to suggest that in the two children with definite clinical illness the culture of staphylococci from the lungs of one and pneumococci from the heart blood of the other was relevant. The child with the positive stool culture for *Sonne* dysentery had been ill, but for the 24 hours before death had seemed well. Other investigators, typically Valdes-Dapena and Hummeller (1963), have found similar lack of help from their further investigations.

Social and Family Background

The social background of the children in both groups is very similar and gives an overall picture of somewhat depressed circumstances. Only four parents were classed in social grade II, none in social grade I, and nearly half the rest were in social grade V. One-third of the children came from families with four or more children. Nearly 10% were illegitimate and a quarter were considered poorly nourished at necropsy. A pin map of the homes shows, to those who know this area, a concentration in the poorer parts of the towns. Fifteen and a half per cent., nearly twice the standard rate, were by definition premature, born weighing less than 5½ lb. (2.5 kg.).

Again these findings agree with those of other observers. Both Coe and Hartman (1960) and Adelson and Kinney (1956) found this high proportion of premature infants (their figures were 15.8% and 14% respectively). The former also found about a third of the deaths occurring in families with four or more children. Both papers report previous cot deaths in the family (in one and two instances respectively). There is less agreement in regard to social background. Coe and Hartman (1960) found on the whole good homes, and infants from all strata of society. Adelson and Kinney (1956) graded homes as poor in 40% and the standard of maternal care poor in 35%; combining the two, two-thirds of the children from poor homes had received poor care. Like Valdes-Dapena *et al.* (1963) they found an undue proportion of negro infants in the series. In this country Elliott (1961) has remarked on the squalor of some of the homes.

Twins

One striking feature of this series is the number of children who were members of a twin pair. Including one dead pair of opposite sex, there were 16 such children, which contrasts with three each in the series of Coe and Hartman (1960) and of Adelson and Kinney (1956). There is no definite pattern apparent with the twin pairs; some survivors were also clearly but mildly ill, but the majority were apparently well. However, in 1959 we had the opportunity of examining a second dead pair of unlike sex twins in a neighbouring town; they were 11 weeks old, born at term, and had developed coughs a week prior to death, apparently recovering with treatment. They were found dead two and a half hours after their last feed and showed the usual typical findings. *H. influenzae* was recovered from the bronchi of the boy.

We have also had the opportunity of examining histological sections and the case records of two further twin pairs from other parts of the country with very similar findings.

Pair C (1962), unlike sex twins, born six weeks prematurely, weight about 3½ lb. (1,590 g.) at birth, were 12 weeks old and apparently thriving when one was found dead in their cot, the other

moribund and dying soon after admission to hospital. The findings were typical.

Pair T (1963), unlike sex twins, 17 weeks old, birth weights 4 lb. 10 oz. and 5 lb. (2,100 g. and 2,270 g.). Found dead together in twin pram four hours after the last feed. Two weeks previously they had had colds which had improved with medicine, only to flare up again with sneezing four days previously. They were active and cheerful, but a little off their food. Apart from numerous blood spots and the other usual typical findings, there were clear pericardial and pleural effusions in both—an unusual feature.

Of the 15 twin pairs in our series 11 were unlike sex twins (including all five in the definite group) as were the other three sets mentioned above. The evidence from twins strongly suggests that infection plays some part in the pathogenesis of the condition, despite the absence of any definite histological evidence.

The frequency with which these tragic deaths occur comes as a surprise to some clinicians, and, indeed, the more the subject is investigated the greater the number of such deaths appears. Swinscow (1951) found that about 600 infants annually were "found dead in bed" in England and Wales and registered as cases of accidental mechanical suffocation. Banks (1958) estimated such deaths as causing over 20% of the mortality in infants and young children, which in 1955 meant over 1,400 in England and Wales. In America Adelson and Kinney (1956) reviewed 126 such deaths of children between the ages of 10 days and 2 years occurring in a two-year period in a population of 1,600,000; Coe and Hartman (1960) found 84 deaths of infants between 5 days and 1 year in an 18-month period in Hennepin County (mainly Minneapolis City), and Valdes-Dapena *et al.* (1963) found 114 in 33 months in the city of Philadelphia (population not stated).

Conclusion

The series presented here started towards the end of 1948, and 120 of the 122 deaths occurred in the 14 years 1949–62 inclusive. The average combined population of the two towns in this period has been a little over 90,000, giving a yearly incidence of nine sudden and unexpected deaths per 100,000 population, and three-quarters of these deaths are in the unexplained or typical group. Furthermore, in the last five years deaths occurring outside hospital and, by their circumstances, reported to the coroner form two-fifths of the mortality in the 2-week to 2-year period, and, perhaps to be expected, four-fifths of the deaths outside hospital.

We consider that our typical group is so uniform as to constitute a disease entity, identical with the cases reported from other parts of the world. In view of the inconclusive and sometimes conflicting evidence of causation, one of us (R. T. C.) has for many years worked on the premise that death has occurred naturally unless evidence to the contrary has been completely convincing, and usually has certified death as due to the non-committal but descriptive "acute congestion of the lungs." We accept the possibility that some children in this group may have died accidentally from suffocation by bed-clothes, etc., but if the overall picture is no different from the group as a whole we do not raise this possibility when reporting, as it cannot be substantiated.

Again, we think that some of the typical group may have died of overwhelming infections, and the twin cases would support this. Young children have poor powers of localizing infection, and the fact that the children in our definite group were older on average may reflect this. But it is difficult to believe that so many children could die of an overwhelming infection and yet have no preceding history of illness, no histological sign of infection, and no pathogenic organism—virus or bacterium—grown.

To us it seems probable that most of the patients in the typical group die from anoxia due to a laryngospasm, arising

either as an anaphylactic reaction to cow's milk (as suggested by Parish *et al.*, 1960) or as part of a generalized neural spasm mediated through the autonomic nervous system (Stowens, 1957) precipitated by the spilling of stomach contents or secretions into the larynx, the irritability of which is possibly increased by hypocalcaemia. Handforth (1959) produced fatal respiratory arrest in rats by a temporary (70 seconds) blockage of the airway. Post-mortem examination showed intrathoracic petechial haemorrhages, pulmonary congestion, and oedema, the changes resembling those found in children dying with "typical" cot death. The presence of mild infection of the upper respiratory tract predisposes to regurgitation; this could explain the preponderance of cases in the overcrowded homes and in the winter months and the preceding history of trivial symptoms. Nevertheless, despite the uniform pattern in our typical group, we realize that no one theory fits all the facts, and the problem remains open.

Summary

Investigation has been made of 122 instances of sudden unexpected death in infancy and young childhood occurring in a fourteen-year period in the Hartlepoons—an annual incidence of 9 per 100,000 population.

In three-quarters of these children certain characteristic findings were present which, however, gave no indication of the cause of death, and in half of this group there was no history of preceding illness. Investigation of the background of these deaths suggests a relation to the season of the year, the social grade of the parents, the birth weight of the child, and the size of the family; there is also a striking incidence in members of a twin pair.

The mechanism of death in the children where this remains unexplained is briefly discussed.

We thank Mr. Norman Graham, H.M. Coroner to Stockton Ward, for permission to publish details of these cases; the Medical Officers of Health for the County of Durham, Hartlepool, and West Hartlepool for information concerning infant life and death in their areas; Dr. A. R. Evans, of Wigan, and Dr. T. A. Harper, of Manchester, for details of their pairs of twins; and the laboratory staff, past and present, at this hospital for their help in these investigations. We have also appreciated helpful correspondence with Dr. R. Ainsworth and the late Dr. A. M. Barrett, of Cambridge, and Dr. J. L. Emery, of Sheffield. We are grateful to Mrs. A. Truefitt for constant secretarial assistance.

REFERENCES

- Adelson, L., and Kinney, E. R. (1956). *Pediatrics*, **17**, 663.
 Banks, A. L. (1958). *Mth. Bull. Minist. Hlth Lab. Serv.*, **17**, 182.
 Barrett, A. M. (1954). In *Recent Advances in Paediatrics*, edited by D. Gairdner, p. 301. Churchill, London.
 Bodian, M., and Heslop, B. (1956). *Eighth International Congress of Paediatrics*, Abstract p. 91.
 Coe, J. I., and Hartman, E. E. (1960). *J. Pediat.*, **56**, 786.
 Elliott, R. I. K. (1961). *Postgrad. med. J.*, **37**, 616.
 Gardner, A. M. N. (1958). *Quart. J. Med.*, **27**, 227.
 Handforth, C. P. (1959). *Canad. med. Ass. J.*, **80**, 872.
 Jones, H. M., and Camps, F. E. (1957). *Practitioner*, **178**, 223.
 Mann, T. P. (1961). *Postgrad. med. J.*, **37**, 620.
 Parish, W. E., Barrett, A. M., Coombs, R. R. A., Gunther, M., and Camps, F. E. (1960). *Lancet*, **2**, 1106.
 Smith, F. B., and Cooke, R. T. (1948). *Ibid.*, **1**, 425.
 Stowens, D. (1957). *Amer. J. Dis. Child.*, **94**, 674.
 Swinscow, D. (1951). *Brit. med. J.*, **2**, 1004.
 Valdes-Dapena, M. A., Eichman, M. F., and Ziskin, L. (1963). *J. Pediat.*, **63**, 290.
 — and Hummeler, K. (1963). *Ibid.*, **63**, 398.

Management of the Chronic Neurological Patient*

DENIS WILLIAMS,† C.B.E., M.D., D.S.C., F.R.C.P.

Brit. med. J., 1964, **2**, 1554–1557

The neurological patient himself rather than his disease is our present subject. In any one family doctor's practice there are not at any one time many purely neurological disorders, but when they do arise they comprise worrying problems of a special nature, they often present a major threat to the patient's future, and they tend to be prolonged. Most acute neurological disorders seen in day-to-day practice are mild, as is for instance the local neuritis caused by the pressure of a disk lesion or by a virus infection, and as is usually vertigo or migraine. Many of the severe disorders may start acutely as a stroke, a fit, or the onset of paralysis, while the subsequent course of the illness may be a slow one, either for better or for worse. Our present concern is for the management of patients with long-lasting disorders which may have had a sudden or gradual onset, and which may improve, may persist unchanged, or may deteriorate. There are, for instance, the cerebrovascular accidents with a long course towards improvement, in contrast to motor neurone disease, where over a couple of years or so

there is remorseless deterioration; there may be the fluctuating course downhill of demyelinating disease, the continuous disability of a congenital hemiplegia or of a chronic traumatic paraplegia, or the episodic course of epilepsy. Not only are the disorders seen to be very varied, but the boundaries of clinical neurology are also very wide, and its application to the patient necessarily involves many other aspects of medicine and of surgery too.

The management of the chronic neurological patient must be based upon the nature of the disorder of function he experiences as well as upon the kind of disease; for though diagnosis is of first importance it is the disability which concerns the patient. Generally, the problems of diagnosis have occupied the physician in hospital, while the family doctor is more occupied with modifying the effects of the consequent disability. Equally, the kind of patient he is trying to help and his attitude to his disability may matter more to the family doctor than the pathological nature of the disease which is causing the chronic handicap. Not only are the patient's personality characteristics which determine his attitude to his disorder important, but his background, which is influenced by the level of his education, by his family milieu, and by his social and economic contacts, contributes for better or for worse to the ease with which he

* From the Pfizer Lecture delivered to the West London Faculty of the College of General Practitioners on 14 April 1964.

† Neurological Physician, St. George's Hospital, London; Physician, the National Hospital, Queen Square, London.