pelvis it is essential that an assistant should disimpact it *per vaginam* before the insertion of stay-suture and incision into the lower segment. Following the incision of the lower segment, the vaginal hand of the assistant pushes the foetal head to a level convenient for the surgeon's hand to take over.

Once the foetal head is stabilized at the desired level in the palm of the surgeon's hand, the position of the anterior ear is established (with 2 fingers of the surgeon's free hand).

(ii) *Correction of asynclitism (aligning head to axis of proposed exit).* If the occiput is oblique or posterior it is easily manoeuvred into the transverse position and asynclitism is realigned favourable to the proposed direction of exit of the foetal head through the uterine and abdominal wall incision. Slight fundal pressure facilitates this procedure.

Knowledge of the exact position of the ear and the establishment of favourable synclitism permits the accurate application of the posterior blade of Wrigley's forceps followed by the application of the anterior blade. Difficulty in the application of the posterior blade will be experienced if the surgeon has failed to disimpact and further elevate the head sufficiently.

If the foetal mouth presents in the uterine incision, as much surrounding blood and liquor as possible is swabbed away and the air passages are cleared with the aid of suction through a rubber catheter. Wrigley's forceps are then applied in this position.

(iii) *Extraction of the head.* Gentle traction on Wrigley's forceps results in the foetal head distending the uterine incision, thereby 'corking' the remaining liquor in the uterine cavity and enabling the surgeon to ensure that the area surrounding the foetal mouth will be dry at the time of egress.

For a number of reasons we regard the surgeon's hand as inferior to forceps for 'delivery' of the foetal head: firstly because the cross-section area of the hand adds considerably to the area which must traverse the uterine incision when the head crowns — consequently greater force is required to achieve delivery; secondly because the further rotation of the hand which finally achieves delivery of the head is the most important cause of tears of an over-distended lower segment (a fact which we have proved beyond doubt in extensive experience of this serious complication); and finally because delivery of the foetal head by means of the surgeon's hand requires the application of an additional force at the uterine fundus which results in the expulsion of remaining liquor — (possibly infected and contaminated with blood or meconium) at the time of delivery of the foetal head. As a consequence the infant's first inspiration may be dangerously contaminated with potentially noxious fluid which could prejudice its chances of survival.

The gentle traction required to extract the foetal head with forceps in a slow controlled manner mimics the ideal natural slow distension of the perineum at vaginal delivery and contrasts strikingly with the forceful — often very forceful — pressure which an assistant not infrequently exerts upon the fundus to achieve delivery when the hand is employed for this purpose.

Extraction of the foetal shoulders and trunk does not constitute a problem.

(iv) *Clearance of the air passages.* Protection of the foetal mouth and nose from contact with liquor, meconium and blood by the interposition of a dry abdominal swab, and urgent clearance of upper respiratory passages with the aid of a size 10 Jacque's catheter attached to suction is an emergency requirement of paramount importance before the infant takes its first breath. Synchronous gentle upward massage of the trachea may aid in the expulsion of potentially noxious meconium if present, as may gentle compression of the foetal thorax after delivery of the shoulders.

A second aspiration of the foetal respiratory passages should follow delivery of the trunk (while the foetus lies head-dependent and semi-prone on the operating towels) before handing the infant over to the staff responsible for its further care.

Breech Delivery

The vaginal delivery of a breech as opposed to a vertex presentation entails additional risks. These risks are reflected — in some measure — when delivery is by lower segment caesarean section. Thus the risks of cerebral haemorrhage, premature aspiration of noxious contaminants of liquor amnii, and difficulty in delivering the after-coming head, are complications which must be fully appreciated and averted in the safe delivery of a breech by lower segment caesarean section:

(*i*) The breech is extracted up to the shoulders by traction upon the foetal feet.

(*ii*) The anterior followed by the posterior shoulder is delivered by a combination of appropriate inclination of the foetal trunk and flexion and adduction of the infant's humerus across its chest, 1 or 2 fingers of the surgeon's hand being employed for this manoeuvre.

(*iii*) The safest method of delivering the after-coming head of a breech is with forceps. The manual method involving more force, mis-directed force, more stimulation to the infant and less control of escape of the head through the incision, predisposes to cerebral haemorrhage, premature aspiration of liquor amnii and of possible noxious contaminants. The head should be brought through the incision occipito-lateral and — particularly in the case of a premature infant — the incision should be adequate, to avoid undue cranial compression.

The anlage of the incision should be maintained as dry as possible with swabs during egress of the head in order to avoid aspiration of liquor and contaminants, and emphasis should be laid upon the immediate suction-clearance of the infant's upper respiratory passages once its mouth becomes visible rather than upon the rapid extraction of the foetal head.

Transverse Lie

(*i*) When the liquor amnii is plentiful and the membranes intact, it may be possible to convert a transverse lie into a vertex presentation before incising the lower segment.

(*ii*) When the foetus is slightly less mobile within the uterine cavity and the foetal back is not posterior, it may be possible to convert a transverse lie into a vertex presen-

SCLEREMA NEONATORUM: A CLINICAL STUDY OF 79 CASES*

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The high mortality[1-3] and the inadequacy of current therapy mirror the lack of data on the aetiology, pathogenesis and course of sclerema neonatorum. In the past sclerema has variously been described as a clinical entity,[1] as a sign found in some cases of neonatal cold injury,[3] or as preagonal induration in severe illness.[4,5]

From a clinical and pathological study of a large group of infants with sclerema we propose the concept that the above descriptions all refer to various stages in the pathogenesis of sclerema.

MATERIAL AND METHODS

A prospective and partly retrospective study of 79 neonates with sclerema neonatorum was made during 1961 - 1963. The cases were judged to have had sclerema in accordance with the definition given by Hughes and Hammond.[1] Severe sclerema was considered to be present when 3 or more sites were extensively involved. Of the 79 infants, 57 were born in the surrounding district and 22 in hospital.

The following data were extracted wherever possible: sex; weight; age and rectal temperature on admission, and temperatures at or shortly preceding death; day of onset of sclerema; its duration and postmortem findings. Other clinical features noted were the occurrence of bleeding, cyanosis, jaundice, oedema, the presence of infection and congenital anomalies.

Special investigations included blood urea and electrolyte concentrations in 35 cases, blood-sugar levels in 7 cases (modified Folin and Wu[6]). A full coagulation study was done in 1 case.

RESULTS

Reasons for Admission

The main complaints and reasons for referral are shown in Table I. Often more than 1 factor constituted the main complaint. Prematurity (45·6%) and hypothermia (20·3%) were the most frequent reasons for admission.

TABLE I. MAIN COMPLAINTS ON ADMISSION AND REASONS FOR REFERRAL (79 CASES)

	No.	*Percentage*
Prematurity	36	45·6
Hypothermia	16	20·3
Feeding problems	11	13·9
Diarrhoea and/or vomiting	10	12·7
Breathing difficulties	8	10·1
Swelling of eyelids or limbs	5	6·4
Weakness or rigidity of limbs	4	5·1
Oliguria	3	3·8
Cerebral birth trauma	2	2·5
Jaundice	1	1·3
Bloody nasal discharge	1	1·3
Exomphalos	1	1·3
Pallor	1	1·3
Maternal intra-uterine infection	1	1·3

Birth Place

The mortality rates whether born in the surrounding district or in hospital were similar (63·5 and 60·9% respectively) and the incidence of severe sclerema in either group was much the same, viz. 67·3 and 69·3% respectively.

Correlations

There were 46 males (58·2%) and 33 females (41·8%), with mortality rates of 76 and 73·1% respectively in the group with extensive sclerema. The mortality was also contrasted with the admission weight (Table II). The majority (74) were premature; only 5 weighed more than 5 lb. 8 oz. (2,500 G). A mortality rate of 65·2% was found in the

*Based on a paper presented at the South African Paediatric Association Congress, October 1964.

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babies weighing less than 3 lb. 8 oz. (1,590 G), while 53·1% died in the group weighing 3 lb. 9 oz. - 5 lb. 8 oz. (1,615 - 2,495 G).

TABLE II. ADMISSION WEIGHT AND MORTALITY RATE

Weight	*Number*	*Deaths*
<2 lb. 8 oz. (1,134 G)	12	10
2 lb. 9 oz.–3 lb. 8 oz. (1,152–1,588 G)	34	20
Total	46	30 (65·2%)
3 lb. 9 oz.–4 lb. 8 oz. (1,616–2,041 G)	15	9
4 lb. 9 oz.–5 lb. 8 oz. (2,069–2,495 G)	12	4
>5 lb. 8 oz. (>2,495 G)	5	4
Total	32	17 (53·1%)
Grand total	78*	47*

*Weight of one infant not known.

Of the 74 infants, 51 (64·5%) were less than 48 hours old on admission, the eldest being 16 days.

Correlating the admission temperature with mortality (Table III) it was noted that 14 of the 17 infants (84%)

TABLE III. ADMISSION TEMPERATURE AND MORTALITY RATE

Rectal temperature	*Number*	*Deaths*
<85°F (<29·4°C)	5	5
85°–90°F (29·4°–32·2°C)	12	9
91°–94°F (32·8°–34·4°C)	16	5
>95°F (>35°C)	12	7
Not recorded	34*	22
Total	79	48 (60·8%)

*23 of these patients were clinically hypothermic.

with severe hypothermia (temperatures < 90°F) died, while only 12 of 28 (42%) with temperatures > 90°F died. Of the 17 temperatures taken shortly before death, 15 were found to be in the normal range. No correlation existed between the rate of rewarming and the subsequent mortality.

Table IV relates the extent of sclerema to the admission temperature. Except for 1 case, all babies with admission temperatures less than 90°F developed severe sclerema, while 50% of the infants with rectal temperatures greater than 90°F manifested with severe sclerema. Comparison

TABLE IV. ADMISSION TEMPERATURES CORRELATED WITH THE EXTENT OF SCLEREMA

Rectal temperature	*Severe sclerema*	*Mild sclerema*	*Total*
<90°F (32·2°C)	13 (91%)	1 (9%)	14
>90°F (32·2°C)	15 (50%)	15 (50%)	30

was made between the extent of sclerema and admission weight (Table V), but no significant correlation was found.

The severity of sclerema correlated closely with the mortality. There were 51 severe and 24 mild cases with mortalities of 74·5 and 28% respectively (the extent of sclerema was unknown in 4 cases). The over-all mortality was 60·8%, compared to an over-all mortality of 22·1% in 960 premature infants admitted to the premature unit in 1963. In the survivors, the maximum duration of sclerema was 6 days.

TABLE V. CORRELATION OF EXTENT OF SCLEREMA WITH ADMISSION WEIGHT

Weights	*Severe sclerema*	*Mild sclerema*	*Total*
<2 lb. 8 oz. (1,134 G)	5 (50%)	5 (50%)	10
2 lb. 9 oz.–3 lb. 8 oz. (1,152–1,588 G)	22 (68%)	10 (32%)	32
3 lb. 9 oz.–4 lb. 8 oz. (1,616–2,041 G)	10 (67%)	5 (33%)	15
4 lb. 9 oz.–5 lb. 8 oz. (2,069–2,495 G)	8 (80%)	2 (20%)	10
>5 lb. 8 oz. (2,495 G)	5 (70%)	2 (30%)	7

Severity of sclerema unknown in 4 cases and weight unknown in one.

Thirteen patients with severe sclerema survived. There were 7 males and 6 females, all of whom had admission temperatures greater than 90°F, and 7 weighed more than 3 lb. 9 oz. (1,615 G).

Haemorrhage

Fifteen cases (18·9%) exhibited haemorrhagic phenomena, and of these 13 died. Sites of haemorrhage included the mouth, nose, rectum, skin, and in 1 case bloodstained oral froth suggested haemoptysis. The 2 survivors had skin petechiae only. Severe sclerema was noted in 13 of the 15 cases. In 2 additional infants, with haemoglobin levels of 10·1 and 11·0 G/100 ml. respectively, intracranial haemorrhage was found at postmortem examination.

Oedema-Cyanosis-Jaundice

Oedema preceding the onset of sclerema, e.g. puffy eyes, swelling of the lower extremities and/or body, occurred in 35 cases (44·3%). Severe sclerema developed in 28 (77·8%) of the 36 infants with cyanosis but only in 18 (52·9%) of 34 babies without cyanosis.

Jaundice occurred with no greater frequency than expected for a normal group of premature infants, viz. in 53·2% of cases.

Complications and Associations

Exchange transfusions were performed in 6 gravely ill jaundiced infants, in 4 of whom sclerema appeared before exchange, while in the 2 others sclerema occurred some 24 hours after exchange transfusion.

Purulent conjunctivitis was present in 4 other cases. Gangrene of the toes occurred in 1 baby, and of the buttocks in another. There were 2 cases with the respiratory distress syndrome, and 1 each of congenital syphilis, duodenal atresia and cerebral birth trauma.

Almost all the infants with sclerema admitted during winter had associated hypothermia. In the 16 infants admitted during summer, hypothermia was not a prominent feature. Sclerema was associated with gastroenteritis in 3, hyperbilirubinaemia requiring exchange transfusion in 2, blood transfusions in 2 and in 1 instance each with cerebral birth trauma, cerebral haemorrhage, congenital heart

disease with cardiac failure and caesarean section. There were no specific features in 5 infants.

Therapy

Therapeutic measures including antibiotics and steroids did not influence the mortality.[7] An interesting observation was the sequence of extensive sclerema appearing within 24 hours after intravenous therapy for dehydration. This occurred in 6 infants, of whom 4 died.

Special Investigations

The observation of high urea and potassium concentrations and low CO_2 content in the blood of infants with severe sclerema has been reported elsewhere.[8]

Blood-sugar levels were determined in 7 infants: the values in 5 fatal cases were 25, 26, 30, 75 and 65 mg./100 ml. respectively. Two infants with blood-sugar levels of 35 and 43 mg./100 ml. survived.

A full coagulation study on 1 infant with severe sclerema revealed a deficiency in factor IX.

Postmortems

Autopsies were carried out in 10 cases, of whom 9 had severe sclerema (in 1 the extent of sclerema was not recorded). Evidence of haemorrhage was found in 7, and included pulmonary intra-alveolar haemorrhage in 6 and intracranial haemorrhage in 2. Three infants had evidence of bronchopneumonia. Sections of scleremato us skin and subcutaneous tissue showed no significant change.

DISCUSSION

In this study a high incidence of prematurity has been observed, and is markedly at variance with previous reports.[1,3] Male preponderance was again noted.

The frequency of hypothermia (70·8%) on admission was striking. Unquestionably prevention of exposure to cold would drastically reduce the incidence of sclerema and cold injury. Infection was an insignificant aetiological factor in this series.

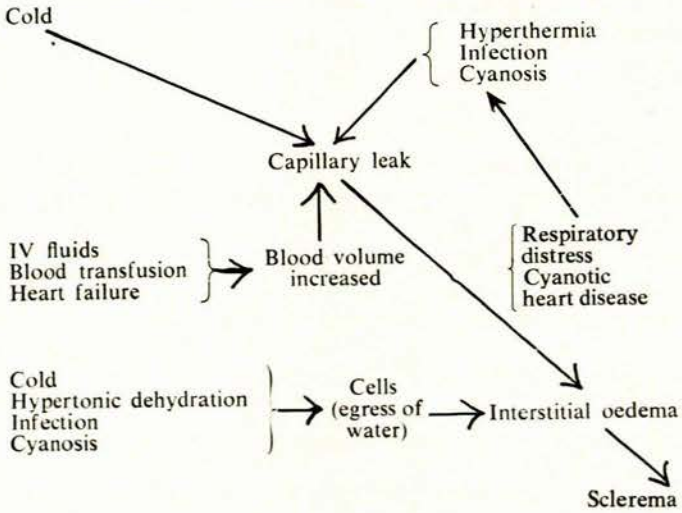
Survival was closely related to admission weight and temperature, i.e. those infants with a greater birthweight and with a normal temperature had a better chance of survival. The prognosis was adversely affected by the extent of sclerema, while the appearance of haemorrhage was an extremely ominous sign. Our impression is that the occurrence of bleeding is closely related to a low admission temperature. Haemorrhage occurred almost exclusively in the severe cases of sclerema. A high incidence of intra-alveolar haemorrhage in the lungs was again observed.[2,3] Its occurrence remains unexplained.

Experimentally, total body cooling in dogs is associated with marked disturbances of *in vitro* clotting tests, and frequently with the development of frank haemorrhage.[9] The additive effects of hypoxia and cold exposure may aggravate the increased vascular permeability which is known to exist in premature infants. A full coagulation study in one of our cases showed a depression of factor IX, a defect which can occur normally in neonates.[10]

Mann and Elliott[2] remarked upon the ease of differentiation between those infants with cold injury and those with sclerema neonatorum, adding that 'it should be apparent that the two conditions have little in common'. However, in the same paper they draw attention to the occurrence of sclerema in some of their cases of neonatal cold injury! Bower *et al.*,[3] reporting 70 cases of neonatal cold injury, found that 39% showed sclerema and stated they 'were surprised to see that it was almost as common in those with relatively high temperatures as in those with low ones'. Notwithstanding their common association, no previous attempt has been made to relate sclerema and cold injury.

We advance the hypothesis seen schematically in Table VI, that neonatal cold injury and sclerema neonatorum are but differing signs with a common pathogenesis. The major factor is exposure to cold, which causes a sluggish circulation and peripheral vasoconstriction. Both in turn predispose to the development of tissue anoxia and hence increased capillary permeability, explaining the appearance of oedema. This constitutes the classical clinical picture of the cold, often immobile neonate, with florid facies and peripheral oedema.

TABLE VI. SCHEMATIC REPRESENTATION OF THE PATHOGENESIS OF NEONATAL COLD INJURY AND SCLEREMA NEONATORUM



Should the hypothermia be sufficiently severe or prolonged, sclerema would appear. The additional operative feature is probably an egress of fluid from the cells into the extracellular spaces as supported by the experimental observations of McCance and Widdowson.[11] It is suggested that the summation of fluid volumes entering the interstitial compartment from both cellular and vascular spaces, causes the sclerema-like feel of the skin and subcutaneous tissues.

Such a course of events would occur with greater facility in premature infants, whose relatively high plasma volumes[12] explain the ease with which their interstitial space may be compromised. The high incidence (93·7%) of prematurity in this series bears out this premise. Perhaps the relatively small interstitial space explains the fact that sclerema is virtually confined to the newborn.

Probably operating by a similar mechanism, is the occurrence of gross sclerema following intravenous fluid therapy, blood transfusion, and cardiac failure, in both this and other studies.[1]

Observations by Mann and Elliott[2] and Bower *et al.*[3] correspond with our experience, viz. that sclerema may appear after accidental hyperthermia or after rewarming a previously chilled neonate. Perhaps supervening vasodilatation increases capillary permeability with resulting overload of the interstitial space. Infection as well as hypoxia of respiratory or cardiac origin may similarly affect vascular permeability causing interstitial oedema. This 'fluid hypothesis' is strengthened by the normal histological features of sclerematous tissue noted in this study and also by Potter[13] and Stowens.[14]

The pathogenesis of sclerema neonatorum is insufficiently explained on the basis of dehydration alone, or on the theory of a shock-like state with peripheral vasoconstriction and the subsequent solidification of the subcutaneous fat.[1] These theories fail to explain the development of sclerema without hypothermia, and its appearance after rewarming a previously chilled infant, the definite role of hypervolaemia, the presence of sclerema and oedema in the same infant, and the high incidence of prematurity.

We feel that our hypothesis is more consistent with the clinical picture as described.

In this study, as in others, treatment was singularly ineffective. In a controlled trial corticosteroids were shown to be of no value.[7] Rational therapy will only emerge when the pathogenesis of sclerema and the exact reasons for demise are established.

SUMMARY AND CONCLUSIONS

During the years 1961 - 1963 inclusive, 79 neonates with sclerema neonatorum were studied. The frequency of hypothermia (70·8%) implicates it as a major aetiological factor in this series, while the high incidence of prematurity (93·7%) was markedly at variance with previous studies. Severe sclerema was observed in 51 and mild sclerema in 24 babies with mortality rates of 74·5 and 28% respectively, and an over-all mortality rate of 60·8%.

The lower the admission temperature, the more extensive was the sclerema and the higher the mortality rate, while the higher the admission weight the lower the mortality rate. The extent of sclerema could not be related to the admission weight.

The occurrence of haemorrhage was almost always associated with a fatal outcome, and the most constant feature in 10 autopsies was evidence of pulmonary intra-alveolar haemorrhage. A common pathogenesis for the development of neonatal cold injury and sclerema neonatorum is postulated.

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