

On Sclerema Neonatorum

A Thesis

submitted

for the degree of

Medicinae Doctor (Glasg.)

by

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SCLEREMA NEONATORUM.

Introduction.

In the conditions, or group of conditions, commonly associated under the name "Sclerema neonatorum", there are found some features of special interest, inasmuch as the disease is one of comparative rarity, and also, as very little as to the causation is yet definitely known.

Although it has attracted the attention of many observers, the conflicting views have tended rather to perplexity than to a clear picture of a well defined disease. This want of definition is reflected in the multiplicity of synonyms whereby various writers have sought to describe the disease. From the time that Usembezius published the first known report, many descriptive terms have been added to the nomenclature. Those which for a time, received recognition include the following ~~the following~~ *List* :—

~~List~~ :— Sclerodermia neonatorum; Scléremie des enfants; Oedématisée concrète; Thirial's disease; Algor progressivus; Induratio telae cellularis; Induratio telae cellularis neonatorum; Algidité progressive; L'engourdissement athrepsique; Sclerom der neugeborenen;

Synonyms

Mal /

Mal de mâchoire; Trismus des nouveau-nés; Hidebound; Scleroma; Sclerisma; Scleremia; Oedematia; Stegnosis; Chorionitis; Scleroedema. In later years, this complexity of titles has tended to simplify itself to the recognition of two allied conditions, viz. Sclerema neonatorum and Oedema neonatorum, with possibly the existence of a third state, the so-called Pseudosclerema neonatorum.

My attention was specially drawn to this disease from having the opportunity to observe a case occurring in my own practice. In this instance, while I consider that the diagnosis of sclerema neonatorum was established, there were absent some of the features usually attendant, such as immaturity at birth, obvious congenital debility or unfavourable circumstances of climatic or domestic conditions.

On reference to various text books (both those on diseases of the skin and those on diseases of infancy) it is notable that most writers dismiss the condition briefly. There is a general unanimity in describing Sclerema neonatorum as a disease found to be present at or shortly after birth, usually in weakly infants, most commonly in those born in unfavourable surroundings; that the disease is characterised by progressive induration /

induration of the skin and subjacent tissues and that, with subnormal temperature and slow heart, it usually terminates fatally within a few days.

In the case which came under my observation, the first indications of disease were noticed thirteen hours after birth, the condition was fully developed within seventy two hours, the induration was almost general over the body and it corresponded in its main features with the published descriptions. In regard to the circumstances of a well developed child in a favourable environment and also in a successful issue the case did not agree with the usual reports, and for these reasons I decided to add my contribution to the subject.

Historical Outline.

Historical
outline

The most satisfactory account of the earliest recognition of this disease was given by Parrot of Paris in 1877 in his lectures on 'L'Athrepsie' at the Clinique des nouveaunés. It is to him that we are indebted for pointing out the distinction between two conditions somewhat allied and, until then, frequently confused, viz. 'Sclerema' and 'Oedema' neonatorum, and it is from his account that I have drawn most of my knowledge of the historical aspect.

The first recorded case was that of a child born in the /

the Stockholm Hospital in 1718; it was said to have been born prematurely and to have died six hours after birth. It was recorded by Usembezius of Ulm in 'Ephemerides Acad. Caesareo-Leopoldinae naturae curios'. (December 1718) under the title of 'Partus Octimestris Foetus Vivus, Frigidus et Rigidus'. This was evidently an instance of congenital sclerema, and Parrot refers to the writer's artless suggestion that it was due to maternal impression, as the mother, during her pregnancy, had spent much time contemplating ecclesiastical statues! In 1732 reference to this case was also made by Schuringius in his 'Embryology' (De Foetu frigido et rigido) and he described the infant as being "from head to foot like a piece of smoke-dried meat". This interesting observation was quoted more than once, but the affection to which it referred was almost completely ignored for many years until Denman, Professor of Midwifery at the Middlesex Hospital, directed attention to it in his lectures, and this formed a starting point for work by Underwood of the London Lying-in-Hospital. In 1770 Underwood published his 'Treatise on the Diseases of Children' and therein made the first valuable contribution to the study of this disease. Under the name of "Hidebound disease" it /

Usembezius
or
Usenbenzius

Schurig
or
Schuringius

Denman

Underwood

it there received description, clinically complete and accurate, as an induration of the subcutaneous tissue. The chief points to which he drew attention were the following:- The disease is usually found in the children of poor people, particularly those in the later stages of an obstinate intestinal ailment where the excreta are like wax or clay; it is not usually found immediately after birth but generally appears within ten days; the first indication is in the alteration in the colour and consistency of the skin, which becomes like soft wax; then the tissues become hard and resistant to the touch but without oedematous pitting on pressure; thereafter there is progressive increase of the hardness, both in superficial area and in density so that the skin cannot be pinched up or slid over the subjacent muscles; even the sheaths and fibres of the muscles may be affected, the rigidity being most marked on the face and extremities but the muscles of the lower jaw are the only ones which become completely fixed; if convulsions occur they do not affect the extremities; the child is always cold, and there is a characteristic moaning cry, often feeble, and quite unlike the cry of an ordinary child of the same age; though he may survive/

survive for some days longer, he seems constantly to be at the point of death; on autopsy, the author had never found any fluid in the cellular tissue; he considered the proximate cause of disease to be a spasm of the skin, and the remote cause an unwholesome atmosphere.

Then arose confusion. Andry of Paris read Under-wood's description and thought it referred to a condition of which he had some cases in the Hospice des Enfants Trouvés. To this affection which, from its supposed incurability, had received little attention, and which actually was Oedema neonatorum, he gave the name "Sclérème," and published his account of it.

In this he stated that the extremities, especially the thighs, are increased in size from infiltration in the cellular tissue; the soles of the feet and the public region are swollen, red and hard: suppuration might, rarely, occur, mortification more frequently, and that, on autopsy, serous effusion is always to be found in the cellular tissue, the fluid being coagulable by heat. Though Underwood recognised the clinical differences between his own cases and those described by Andry, it was supposed that the variation of type might be due to climatic conditions in London differing from those in Paris.

Andry's /

Andry

Andry's error was repeated by others and contributions by Capuron, Auvity, Chambon and Léger all perpetuated the confusion between sclerema and oedema and they united in including the oedema under the name of "Endurcissement du tissu cellulaire." A contribution by Denis would suggest that he had, in some measure, appreciated the error of previous writers, inasmuch as he sub-divided the 'endurcissement du tissu cellulaire' into two varieties, serous or oedematous and fatty or solid, but his further conclusions did not assist progress. He considered that the two varieties presented the strongest analogy in respect to their symptoms, their progress and their termination, while the differences were unimportant in the colour of the skin, the consistency of the tissues, and the fact that the cellular tissue in the one case was infiltrated with serum, and in the other laden with fat.

Denis

Billard recognised two varieties of induration, and gave a clear description of oedema neonatorum but he considered the true sclerema as a condition found after death, or, at the earliest, in infants actually at the point of death.

Billard

Valleix spoke of 'fatty induration' as being entirely different from the oedema, but, along with Billard, /

Valleix

Billard, he considered the former to be a post mortem change.

Bouchut added another complication by insisting that this condition is not peculiar to the new born, that it is in fact, the same disease as scleroderma. According to him, it is unjustifiable to differentiate between the two varieties, simple and oedematous, the oedema being only a consequence of the primary induration; although he claimed that the induration was the essential lesion, he gave no detailed description of it, stating only that it was necessarily a condition of the moribund state. In brief, while attempting to describe 'Sclérème or induration of the skin of the new born', Bouchut really described a non-existent type of disease.

Bouchut

In 1873 Clementowski of Moscow published a contribution (Oesterr, Jahrbuch für Paediatr.) in which he distinguished three varieties of Sclerema viz:- the erysipelatous, the oedematous, and the adipose. The first referred to the subcutaneous serous infiltration often found with erysipelas of the new born; the second was oedema neonatorum; and his description of the third was applicable quite as much to cadaveric rigidity as to any type of disease.

Clementowski

I have quoted points from some authors to indicate the utter confusion which, for many years, blinded many observers in the actual recognition of the disease. Andry's original error in applying Underwood's description of one affection to another and distinct morbid condition soon received academic recognition, and his example was followed by other writers not only those above mentioned, but by those in Britain - Coley, West and others - and by most of the German authorities. This chaotic state continued until Parrot emphasised the clinical and pathological lines of demarcation between the two maladies and, for the first time, offered a rational appreciation of the situation.

His example was followed by Henoch of Berlin, who, in 1881, in his lectures on diseases of children, differentiated clearly between sclerema and oedema. That this recognition did not receive universal acceptance is demonstrated by the fact that, for years afterwards, in various standard works, even in those devoted to diseases of children, the writers were content to dismiss these ailments without notice, or to describe them in such a manner as to shew that the confusion still existed. The fullest account in English is that by Ballantyne, while contributions by other /

Henoch

other observers - Barrs, Langmead, Money, Mackenzie and others - have, for the most part, been in the form of extended notes of individual cases.

General considerations.

Reference has been made to the fact that, amid the various opinions as to the exact nature of Sclerema there is some consensus of opinion as to its mode of origin, although the generally accepted facts do not carry us very far in our knowledge. It has been recognised that the disease is most likely to be found where there are general conditions inimical to health. Immaturity at birth, congenital weakness, congenital syphilis, illegitimacy, parental poverty or maternal debility and also the occurrence of birth during very cold weather, have all been taken as factors in individual cases, but we are still without positive knowledge of the exact cause. Ballantyne (B.M.J. 1890) holds that there is no proof of direct association with congenital syphilis, but he claims that "probably the diseased process is initiated in utero." - It may be mentioned incidentally that in the case which I have to report samples of blood from the child's parents were examined for me, the Wassermann test in each case being negative.

General
considerations

To /

To give some explanation of this comparative ignorance of a much discussed condition, three points may be mentioned. One is the actual rarity of the disease (although Osler suggests that it may be much more common than statistics would indicate) so that few opportunities are afforded for its study. The second point is that, even when the disease does occur, it may probably be in surroundings which are not favourable for careful examination by any competent observer. A disease which is so commonly and so soon fatal, occurring in a premature or illegitimate infant, perhaps born into poverty-stricken or squalid surroundings or amidst adverse climatic conditions, might easily fail to be recognised; death would be ascribed to the other obvious disabilities, and the brawny induration might be noticed only after death, and be then supposed to be post mortem rigidity. Further, it is also probable that mild cases may occur and may become well without being recognised. Garrod refers to the occasional observation at out-patient dispensaries, of infants, generally of a few weeks old, having localised areas of induration which has all the characters assigned to sclerema. In these cases, the prominent symptoms have passed off, and he (Garrod) has had to watch only the /

the gradual resolution of the sclerematous lesions, a process usually lasting five or six months. He considers that such cases, if true sclerema, are less rare than the fatal cases.

General
considerations

In this limitation of our knowledge, it is impossible to give a comprehensive definition of the disease, its cause, its variations and its characteristics. Ballantyne offers a provisional definition which may be taken as reasonable, viz. "a rare disease occurring most commonly in the new-born, characterised by induration of the subcutaneous tissue, and little amenable to treatment" (B.M.J. 1890) He expresses the opinion that, without fuller knowledge of the pathological and physiological processes concerned, a more scientific definition is impossible.

Parrot states that sclerema is one of the signs of athrepsia, and that athrepsia is only a complication of oedema neonatorum; this opinion, as I hope to shew, does not seem to be justified. He says "the induration under the influence of athrepsia --- takes possession of these parts for which it has affinity and expels from them the serous fluid with which it cannot co-exist. One cannot give a better proof of the incompatibility of the two affections. One displaces the other because they can never exhibit themselves together in the same situation." Reference will /

will be made later to a striking example quoted by him where the two conditions, sclerema and oedema, were found in the same case, though not simultaneously.

General
considerations

In his clear differentiation between the two conditions, 'sclerema' and 'oedema' of the new born, Parrot gives a graphic picture of the former condition, and it seems fitting, although the quotation is somewhat lengthy, to give here some points of his description, as an amplification of the brief account given by most other writers.

"When the disease (athrepsia) develops slowly, from Parrot
"there may be two very different results which I wish
you to recognise without its being possible for me
"to state precisely all the circumstances which deter-
"mine the one rather than the other. In the one case ---
"In reference to the other condition it constitutes
"the athreptic induration. We notice this specially
"when the disease assumes a subacute form almost
"immediately after birth, in subjects of moderate
"plumpness. The skin, far from forming folds, becomes
"on the contrary stretched and its surface becomes
"remarkably smooth; it loses all flexibility, and it
"is absolutely impossible to separate it from the sub-
"jacent parts, with which it seems to be very closely
"united. This alteration begins in the lower limbs;
"the lumbar region is then invaded, then the lower
"part /

"part of the trunk and finally the whole body, the
 "face included. Daily one may see the tension and
 "the hardness of the skin make progress, and soon on
 "touching it, one has the feeling which thick leather
 "gives. It appears as if all the soft parts were
 "coagulated, and as if one had before his eyes a
 "figure of wood or marble; Therefore the first
 "observer who saw an infant affected in this way
 "imagined guilelessly that the mother had gazed at a
 "statue. The integument is not to be pitted by
 "pressure of the finger, and its colour becomes faint-
 "ly bluish or livid. Immobilised by this rigid con-
 "dition, which cannot be overcome spontaneously, the
 "limbs remain extended, and, but for certain move-
 "ments of the thorax and face which one still observes,
 "one might believe that the body is in a state of
 "cadaveric rigidity. Dugés reports that on grasping
 below the head infants thus affected he was able to
 hold them in a horizontal position, as if they had been
 made in one piece only. It has often happened for me
 "to make the same experiment in another fashion; by
 "applying the radial border of my hand under the back
 "of the little invalid, I have held him up as if he had
 "been cast as a rigid shaft. When the face is invaded
 "the stiffness of the ^{lips} ~~limbs~~, of the sides of the cheeks
 "and /

General
 considerations
 from Parrot

"and of the muscles, keeps the mouth closed and
 "renders sucking and swallowing impossible. Thus one
 "understands how a certain number of observers from
 "the time of Lodmann quoted by Denis, until our own
 "day have believed that they had to deal with the
 "affection called 'lockjaw' or 'trismus of the new
 "born' -----

General
 considerations
 from Parrot

This induration of the new born,
 where the muscles and the cellular and fatty tissues
 "are all affected as much as the skin, is invariably
 "the result of athrepsia. I have never seen this
 "develop apart from it. -----

"In the latter days of athrepsia, the peripheral
 "soft parts diminish, dry up and harden; the skin
 "becomes stretched and assumes a livid colour; the
 "whole body becomes rigid, immobile and seems mummi-
 "fied. This characteristic condition is only met
 with in Athrepsia and hereafter I shall call it by the
 "name of 'endurcissement athrepsique des nouveau-nés'.----

"Of the posture I have only a few points to mention
 "to you ----- the state of the limbs is generally as
 "follows:- the thighs are drawn up to the trunk and
 "the legs are bent upon them: the toes are sharply
 "bent towards the sole of the foot as if there were
 an actual contracture: the arms and forearms are
 "stretched /

"stretched but the wrists are flexed on the latter
 "and the fingers bent into the palm of the hand about
 "the thumb. One must use some force to overcome the
 "flexion, and when one has straightened out these
 "parts, one sees them quickly return to their former
 "position. These movements, furthermore, provoke
 "but little pain, and some infants bear them without
 "arousing from their stupor.

General
 considerations
 from Parrot

"The skull undergoes some important modifications.
 "Its size becomes less, as is proved by the condition
 "of the osseous portions which form the vault, and by
 "that of the soft parts which unite them. The project-
 "ion which the fontanelle causes in health, gradually
 "sinks down, and in its place one may see an actual
 hollow, the depth of which may reach three or even
 "four millimetres. The diameter of this membranous
 "surface also decreases in a very notable fashion by
 "the approach of the bones which surround it, and in
 "some patients it disappears almost altogether.

"At the same time as M. Bouchaud was the first to
 "observe, the sutures become fixed by the disappearance
 "of the interosseous spaces. The bones first draw
 "together, then over-ride, in such a manner as to
 "form linear projections. "These, always appreciable
 "to /

"to the touch, can also be proved by sight in many
 "cases where the phenomena, well marked, become more
 "obvious by the thinning of the hairy scalp and by
 "its stretching over the calvareum,-----

General
 considerations
 from Parrot

"When the induration extends also to the face, one
 "might say that a stiff mask covered it and there is
 "no visible movement. The jaws, clenched on one
 "another cannot be opened without force, and if one
 "happens to separate them, they come together again
 "as if moved by a spring. Add to this the pained
 "expression and you will have all the characteristics
 "of the athreptic physiognomy" -----

I have thought it of value to translate and
 incorporate these portions of Parrot's description of
 sclerema. They are not given consecutively by him,
 and I have omitted parts which seemed applicable to
 his conception of 'athrepsia' in general rather than
 to sclerema in particular, but with his acceptance
 of Underwood's description, they serve to illustrate
 some of the appearances which I have not seen described
 elsewhere. In another place, Parrot quotes a des-
 cription by Denis which also seems worthy of repetition.
 "The indurated parts of the body are neither elastic nor
 "doughy; from the effect of the disease they have
 "acquired the firmness of suet. The skin, of a
 "yellowish /

from Denis

"yellowish white colour, has the appearance of
 "leather; it does not slip over the subjacent
 "muscles, and one may feel under it, here and there
 "very firm masses. When the subject is in the third
 "stage of induration one might believe that it had
 "undergone freezing; on percussion, the limbs some-
 "times give the sound of wood. The coldness is in-
 tense, the movements difficult; trismus and the
 "other nervous phenomena are less common than in the
 "oedematous variety; the respiration, the circu-
 "lation and the digestion are also less disturbed.
 "All these phenomena are attributed to the accumu-
 "lation of the cellular fat in the vesicles, and
 "to the increase of its density." -----

General
 considerations
 from Denis

The following report from Parrot (L'Athrepsie
 p.128) is quoted by him to illustrate his conten-
 tion that 'sclerema' and 'oedema' are conditions
 "which "are not only different but, so to say,
 "opposed"; it is also of interest as shewing the
 "progressive diminution of weight.

"The following observation is a most striking
 "example of the difference and of the incompatibility
 "which exist between athreptic induration and the
 "oedema of the new born, because it demonstrates them
 "both /

from Parrot

"both to us in the same case but at opposite ends of
"the disease.

Case reported
by Parrot

"Case V. Anasarea: athrepsia: absorption of effusion;
"athreptic induration: - pneumonia.

"Florence D. born 27th October 1875, admitted to the
"infirmary 30th. The skin is red on the different
"parts of the body except at the extremities where it
"is bluish. There is some oedema most marked at the
"hypogastrium and at the labia majora. The infant is
"passing meconium, and the cry is feeble. Weight 2063.
Pulse 100. Temperature Ax. 32.2° 31st Temp. Ax. 37.2°
Rect. 32° Rect. 37.2°

"1st November. The colour of the skin and the oedema
"are decreasing; at present the latter is still well
"marked on the right thigh. Weight 1962.

Ax. 36.4°
"2nd Weight 1900 Temp. Rect. 36.1°

"3rd. The oedema is present only on the labia majora.

"The bones of the skull overlap. The umbilical cord

"has not fallen off. The skin is still red. Weight

Ax. 37°
"1810. Temp. Rect. 36.8°

"4th. The skin has entirely lost its red colour; it
"is pale. Emaciation.

"5th. The infant takes very little milk. Weight 1715.

Ax. 37.4°
"6th. Weight 1673. Temp. Rect. 37.4°

"9th. /

"9th. There is some induration of the soft tissues.

Case reported
by Parrot

"At the back of the right lung one may find some

"crepitant rales, with coarse and noisy bubbles. Pulse

Ax. 34.1°

"132. Weight 1485. Temp. Rect. 34.1°

"10th. The child's skin has the consistency of a

"cuirass and gives the whole body a wooden feeling

"and absolute rigidity. There is almost generalised

"immobility and some trismus. The eyelids are stretch-

"ed and as if pasted on to the eyeballs, and it is

"impossible to open them. In whatever position one

"puts the little patient it lies quite rigid. Pressure

"on the breasts causes a little milk to come away.

"There are some crepitant rales at both sides behind.

"The heart's sounds are slow and dull. Pulse 80. Weight

1460. Temp. Ax. 39.4°
Rect. 30.4°.

"Death took place at 7 p.m. Autopsy on 12th. Weight

"1433. The centre of ossification of the lower end of

"the femur is scarcely so large as that of a mature

"infant. Pleuropneumonia of the lower part of both

"lungs. There is some pus in the ears."-----

Some consideration must be given to the possible concurrence of oedema and sclerema. Parrot's claim that the two conditions could not be co-existent was in accordance with the views held by some of the more accurate clinicians, but in more recent years there has been /

been a tendency to recognise an indurative and an oedematous variety of sclerema. G. Somma recognises three varieties of sclerema viz.- "indurative," "oedematous" and "mixed" according to the state of the subcutaneous tissues, and Ballantyne indicates his acceptance of the statement that oedematous infiltration may occur in sclerema. Finkelstein in the 'Lehrbuch der Säuglingskrankheiten' (1908) says that in some cases sclerema and oedema may be associated together, and the oedema may be so bad that pitting on pressure may scarcely be got, so that diagnosis during life may not be possible.

G. Somma

Finkelstein

I would suggest that in those cases where serous infiltration is found it may be considered, not as an essential feature of sclerema nor characteristic of any special type of the disease, but only as a secondary sign. This idea is corroborated by the fact that cases of sclerema have been found with serous effusion into the peritoneal or pericardial cavities, and in others there has been oedema of the epiglottis and vocal cords.

In the more recent literature to which I have had access I have not found many other points of importance in regard to macroscopic features. Mensi in the 'Rivista /

'Rivista di clinica pediatrica' (1911) had an article on 'Lo Sclerema di neonati' of which an abstract is given in 'Zeitschrift für Kinderheilkunde' (1911). From personal observation of eighty two cases, he recommends the differentiation of sclerema into two types according to the appearance of the skin. The first occurs in premature and newly born infants and in the cold season of the year: it follows a previous oedema, and the induration appears first in the buttocks, never or seldom in the hands and feet, and it spreads more or less over the body; the temperature is usually low, seldom febrile; common complications are bronchopneumonia and nephritis; and the disease usually ends fatally. On autopsy the skin is found to be atrophied, with absence of the granular layer; the cells of the cutis are closely packed together and there is dilatation of the vessels with haemorrhages into the cutis.

The second type is characterised by condensation, thinning and drying of the skin; it occurs less often in premature children, mostly in warm weather and it may begin with oedema; it also begins in the buttocks and may become general. Usual complications are diarrhoea and fever, causing death. On examination post /

post mortem there is found to be atrophy of the dermis, with condensation of the fibres, scantiness of the cells and vessels of the skin, and abnormal development of fibrous tissue round the fat --- Mensi considers that the first type corresponds to Sclerema neonatorum and the second to Sclerema adiposum of Parrot.

In the matter of temperature some very striking records have at times been made, confirming the general observation of subnormal temperature as a common feature in this condition. Henoch makes note of temperature as low as 83.3^o Fahr. Verson records one of 79^o Fahr., while Roger had one case even as low as 72^o Fahr. Depression of the temperature however is not an invariable sign, and in other cases a greater or less degree of pyrexia has been found present, at least for part of the currency of the illness.

Temperature

Another variable manifestation is in the coloration of the skin. 'Ivory pallor' 'dirty yellow' or 'waxen appearance' and different tints of bluish, purplish, or livid colour have been mentioned in the description of some cases, while in others again no change from the normal colour was to be recognised. Henoch reports /

Colour

reports that almost all of his cases were more or less jaundiced.

Other points referred to by some writers are the facts that in sclerema there are not found any of the changes in the urine which are characteristic of oedema and, also, that in sclerema the lax tissues of the eyelids, the scrotum or the labia majora are not liable to be affected and swollen as in oedema.

In the 'Lancet' (July 1906) Carpenter published a report of a successful result in a case under his care in the London North Eastern Hospital for Children. He refers to the rarity of the disease and to its being even less common in England than in France and Italy. He also states that there are "two disorders, differing at any rate in degree, which both go under this name" (i.e. Scl.neonat) The more common type occurs in children, otherwise normal; it is limited to the skin and subjacent tissues, and it usually ends in recovery. The second type attacks weakly or premature children, and it is accompanied by severe disorder of the alimentary, respiratory or renal systems, ending commonly in death.

Carpenter

In the case described by him the child was six weeks old, well developed and apparently normal except for /

for the cutaneous induration on the posterior aspect of the body, the back of the scalp, neck, trunk, buttocks, arms and thighs being all affected. In the regions affected the skin was of a pink-purplish appearance with well marked dimples and slight rounded eminences. On pressure with the finger no pitting occurred but the colour faded to white and it slowly returned when the pressure was removed. These areas ended by gradual thinning and they were not painful. At a later stage the induration extended to the angles of the jaws, the sides of the neck and down the arms, with islets all over the abdomen and chest. Examination of the blood was inconclusive, the relative proportions of the various corpuscles given by him being quite within the normal limits found in infancy. A small piece of skin and subcutaneous tissue was excised from the thigh for examination and, macroscopically, the fat appeared distinctly whiter, much harder and in larger globules than normal. Microscopically, no definite abnormality was found, the serous infiltration and increase of connective tissue, sometimes described, being definitely absent in this case. The gradual progress of the child was slow and irregular and after five months' observation, there were patches of induration still present. /

present.

As a corollary to these general considerations about Sclerema, it is desirable to make some brief reference to some conditions which may reasonably be linked with it, and in which there might be difficulty in the differential diagnosis. Of these, Oedema neonatorum is the most important, if for no other reason than the fact that the two conditions have so frequently been mistaken for each other.

Differential
diagnosis

Oedema neonatorum is to be regarded as a sign of disease rather than as constituting in itself a definite and self-existing condition. It is found usually, if not invariably, in association with other recognisable pathological affections, renal, pulmonary, cardiac or intestinal, such as those which commonly cause oedema in adults, and its distribution in the body is, as one might expect to find it, in those situations where the influence of gravity or the laxity of the tissues would be most favourable to its occurrence.

Oedema
neonatorum

In one case recorded by Ballantyne (B.M.J. 1890) the child (male) was born while the mother was suffering from acute bronchitis with pneumonic patches; within a few hours after birth he developed marked oedema, with suppression of urine and he died in two days; on section there was found tubular and glomerular nephritis; in /

in this case the condition may reasonably be ascribed to the maternal toxæmia. The most important feature distinguishing it from sclerema is the pressure of serous fluid in the interstices of the subcutaneous tissue. This will, in nearly all cases, allow of pitting on pressure, though it is possible that in an extreme case the infiltration may be so firm that pitting may not be got at all.

Oedema
neonatorum

The presence of oedema neonatorum is always to be held as a grave omen and such cases usually end fatally. Kaposi suggests that the proximate cause is the retardation of the capillary circulation, caused, remotely, by systemic disease or by extreme weakness. After death, whatever the pathological findings as to the primary disease, there will always be found the characteristic serous effusion in the subcutaneous tissues, and microscopic sections, which, from the looseness of the tissues, are not easily made, will shew bundles of fat cells, loosely connected by thin bands of fibrous tissue.

Syphilitic roseola might conceivably give trouble in diagnosis from sclerema, but in such a case, apart from the actual appearances, the other facts of the history, the Wassermann reaction and the results of antisyphilitic treatment would serve to establish an opinion. /

Syphilitic
roseola

opinion.

Tetanus neonatorum, another rare condition, might need consideration as a possible alternative to the diagnosis of sclerema. If due to the earliest possible infection, as from inoculation of the raw surface of the umbilical cord, it might develop within eight or nine days. The first indication would probably be trismus, the affection would be indicative of the usual tetanic signs of muscular spasm rather than of subcutaneous induration, the rigidity would be greater than ⁱⁿ ~~the~~ sclerema, and the temperature curve would be different to that found in sclerema.

Tetanus
neonatorum

Erysipelas neonatorum may also arise from umbilical infection or from a mother with puerperal fever.. it usually occurs within ten days after birth, and it presents the features of continuous pyrexia, and the blush with its sharply defined and advancing margin, without any material difference from the usual indications of erysipelas.

Erysipelas
neonatorum

Fat-sclerema is another of the class of symptomatic conditions and it may be found in young infants where there has been marked drainage of fluids from the body, as from severe vomiting or diarrhoea. It occurs after death or, occasionally, just before it, and it is supposed to be due to solidification of the panniculus adiposus.

Fat-sclerema

Description /

Description has also been given of a condition ^{"Pseudo-sclerema"} called "pseudosclerema" but it is open to question whether this ought to be considered as having any essential pathological difference from true sclerema.

The general account of it is that this is a condition resembling sclerema neonatorum but where the signs are less marked and where the disease does not end in death. This description may be paraphrased, not unfairly, by saying "if it is a bad case and the child dies, it is a true sclerema, while if the condition 'is less severe and the child recovers, it is a pseudo 'sclerema."

In the absence of full knowledge and while the aetiology is mainly speculative, it seems to me more reasonable to hold that the two conditions, sclerema and 'pseudo-sclerema' so called, are essentially the same, and that the differences in the signs and the result are dependent on the individual child itself, on its own vitality, and its degree of resistance.

Due regard must be given to the statement of Henoch that infants affected with sclerema "invariably die." One is reluctant to traverse, perhaps on inconclusive grounds, any statement made by one of his wide experience. And yet, such an observation, although it is the honest opinion of a careful clinician, is obviously gained from his own experience, and in regard to a disease /

disease such as the one under review, the limited possibilities of studying the affection must be remembered. It is also pertinent to remember comparable opinions by others of equal eminence in their own spheres, where fuller knowledge has proved their inaccuracy. It would need no laborious 'delving into the musty records of a bygone past' to find competent surgeons who held definitely that the opening of the peritoneal cavity must inevitably and invariably cause death, and now within the space of one generation, laparotomy has become almost a commonplace of surgical procedure.

To the preceding section of this thesis, I have applied the heading 'general considerations', because it includes more than the merely diagnostic features of the disease. From the main points quoted above, it is, in my opinion, reasonable at this stage to draw some general conclusions on the subject, and these may be summarised as follows. Sclerema neonatorum is to be recognised as a well-defined disease, not necessarily associated with or secondary to any other recognisable morbid condition; it may present conditions varying widely in degree as to the extent of the local lesions, the severity of the constitutional symptoms, and the gravity of the ultimate prognosis: further, these conditions will vary inversely as the degree of individual /

Some
conclusions

individual resistance or immunity, and, possibly, may vary directly as the degree of infection by the primary cause: and, finally, the differentiation of cases into true and false types of sclerema is not based on any sound or scientific premises.

Personal observation.

In the two cases of which I have had the opportunity of personal examination, the first is of special interest in regard to the apparently favourable condition of the child, and also in demonstrating that wide-spread development of the induration and severe constitutional symptoms are not incompatible with recovery.

Personal
observation

This case occurred in a household which has been known to me for some years, and the family history on both sides is particularly good. The father is a carpenter, a well built, healthy and sober man, age 39, height 5ft. 10 in., weight 15 st. He comes of a family of farmers and his eight brothers and two sisters are all alive and well. The mother, aged 33 years, is also from a healthy stock; her paternal grandmother is living, her father is in good health, her mother died of puerperal fever and her four brothers and two sisters are all healthy.

The /

The parents of the infant live in a clean and new tenement, with all reasonable comfort of their social position. This child is the third born, and during her pregnancy, the mother was in specially good health.

Personal
observation

Labour began on the afternoon of 18th October, the actual day which I had previously indicated as the probable date of delivery. The pains were slight and infrequent until after 8 o'clock p.m. when three quick and forcible pains completed the second stage spontaneously, the child being born immediately after a messenger had been sent to summon me. The child appeared to be a plump healthy boy, and he weighed 8 lbs. The placenta was examined, and it shewed no abnormality. On the 19th October, about thirteen hours after the birth, the nurse noticed that the child's face was of a dark bluish colour. This cyanosis passed off soon but it recurred in the afternoon, five or six hours later. During the following night (19th-20th October) these attacks came on more frequently and more violently, occurring almost every hour. Each attack began with three sharp screams, the eyes rolled, the hands became rigid /

Personal
observation

rigid with fingers clenched and there was marked cyanosis of the face. Each seizure seemed to last about ten minutes, and it passed off gradually, leaving the child exhausted, but he had no proper sleep till the 22nd October. They continued with about the same frequency until the evening of 21st October.

When I saw the child on the 20th October the report given of these attacks with the marked cyanosis suggested the possibility of cardiac malformation, but I could not detect any cardiac abnormality except the pulse rate which by auscultation I estimated as being about 200 per minute.

On the same day (20th October) the nurse drew my attention to the firmness of the thighs, and on having him undressed, I found the same condition over the thighs, buttocks and dorsal regions. The most notable feature was the half-frozen feeling or rubber-like consistency of the parts on pressure; there was no pitting, even on firm pressure; the skin could not be picked up from the underlying tissues, and, even when freely handled, there did not appear to be any pain. This firmness had been first noticed by the nurse on the afternoon of the 19th October and it was then limited to the buttocks and the legs below the knees. Within /

Personal
observation

Within 72 hours after birth, the induration was found over the dorsal and lumbar regions, also about the shoulders and face and jaws, with patches on the front of the abdomen and in the lateral costal regions. In these areas, the affection seemed to be in broad patches rather than being limited to any special groups of muscles.

The skin appeared to be normal in colour without any of the abnormal tints described by some writers. I spoke of the case to Dr Leonard Findlay, because of his special interest in the diseases of infancy: he saw the child twice with me, the first time being on the 21st October and he agreed with me that the case was one of Sclerema neonatorum. At his first visit, the child seemed to be quite unconscious and the rectal temperature was 97⁰ Fahr.; there was external strabismus of the right eye, but this did not long remain present.

For the first week the child was extremely ill, and I could give very little prospect of recovery. From that reason, I can give no continuous record of temperatures, as I was unwilling to have him disturbed to the extent necessary for taking the temperature in the rectum or axilla, particularly as such a procedure was not required for any purpose of treatment. There was not any noticeable degree of coldness of the skin, but this point must be considered in /

in association with the fact that he was kept almost constantly in an extemporised incubator so as to maintain artificial heat.

Personal
observation

During the first six days there was great difficulty in giving him any nourishment, this being due to the muscular rigidity. He was quite unable to suck, and had difficulty even in swallowing; our only method was to open the mouth and pour in about half a teaspoonful of fluid which would be swallowed slowly. After the sixth day the stiffness left the facial muscles and he became able to swallow with increasing ease.

While the onset of the disease was sharp - the change of appearance from that of a normal healthy new born child to that of an infant apparently at the point of death occurring within seventy two hours - the recovery from the disease was very gradual and without any well-defined stages. Dr Findlay at his second visit (2nd November, 1912) could recognise a marked diminution in the degree of hardness as compared with that found by him twelve days previously; it was also noted then that in the mid dorsal region there was an area of induration which seemed to be painful when handled.

The /

Personal
observation

The progressive disappearance of the stiffness particularly as it affected muscular action, was first noted after six days in the face and jaws. Thereafter, it gradually faded away from the thorax and abdomen, then from the dorsal region, then from the buttocks and shoulders, and lastly from the outer aspect of the thighs. This process of gradual resolution occupied fourteen to fifteen weeks, and on each thigh there was a firm patch easily to be felt for about three weeks after the other areas affected had regained their normal consistency.

After the child was about ten weeks old, there was practically no need for medical attention; he has made steady progress and is now in good health; at 10 months he weighed $15\frac{1}{2}$ lbs nett. and in May, 1914 - *19½ lbs. nett.*

Dr Findlay's
case

Of a second case I am able to give some account by the kindness of Dr Findlay who saw it at the out-patient dispensary of the Glasgow Royal Hospital for Sick Children. I was unable to see the child alive but I had the opportunity to assist at the autopsy. This child was male, illegitimate, born in miserable surroundings, and there was well marked congenital syphilis; the mother stated that there had been a rash on the body when he was born; he had taken the breast freely, and there /

there had been neither vomiting nor diarrhoea. He was supposed to be mature but, when first brought to the dispensary on his twelfth day (9th October 1913), his weight was only 4lbs 10oz. Dr Findlay's note of his examination makes mention that the body was small and emaciated, and there was a pemphigoid eruption over the body, specially on the buttocks, legs and arms, while the soles and palms were also implicated; the rash was well marked round the mouth and there was a patch on the left cheek; the mucous membrane of the lips was excoriated and there was an ulcer on the right anterior pillar of the fauces; the spleen was only just palpable; there was then no sign of sclerema. On the following day (10th October 1913) he was again brought to the dispensary, and it was then noted that there was a well marked and extensive rubber hardness which the mother had first noticed on the morning of the same day, so that she had difficulty in moving the limbs. This sclerematous stiffening affected the face and chest and it was well marked on the buttocks, thighs and calves, also about the deltoids, the arms and extensor aspect of the forearms; there was no oedematous pitting, even on firm pressure. The child was jaundiced and obviously weaker than on the /

the preceding day but the cardiac sounds were comparatively good; he was unable to suck; the rectal temperature was 96⁰ Fahr. Neosalvarsan grm 0.075 was injected into the right gluteal muscles. The child died at 3 a.m. on the following day (11th October 1913) when fourteen days old.

When I assisted Dr Findlay at the post mortem examination (12th October 1913) the following conditions were found ---- Icterus. Dried blebs of pemphigus, well marked on arms and hands, legs and feet, and on the lower abdomen. In the head of the pancreas a gumma about the size of a cherry. Left kidney shewed an abnormally large suprarenal capsule; right kidney seemed normal. Liver was enlarged and of a dark greenish colour; gall bladder could not be emptied by pressure, on account of fibrotic thickening in the portal fissure. Brain was soft and oedematous, and the surface markings were not well defined; there were no signs of haemorrhage nor of thickening of the meninges.

Subsequent examination shewed the presence of spirochaetes in the pancreatic gumma, also in the pancreas, liver and adrenal capsules. Blood from the mother gave a positive reaction to the Wassermann test. Sections of skin from the buttock were prepared by Dr W.B.M. Martin, pathologist to the hospital, and by his /

his kindness, I am able to give photographs of them and also of normal skin from the abdomen of an infant of three months. Comparison of these photographs does not shew any increase of fibrous tissue in the case of sclerema as compared with the normal skin, but in the former there is a marked thickening of the rete Melpighii, and, perhaps some abnormal increase and aggregation of the cells of the adipose tissue immediately underlying the dermis.

Morbid anatomy.

On considering the pathological alterations found in the tissues as a result of sclerema, one finds some degree of unanimity in the more trustworthy descriptions. One important fact which seems to be well established consists in the absence of any constant visceral lesion. In the examination of such cases after death, there have been found at times various changes in the internal organs. The most notable of these have included pulmonary affections such as atelectasis, pneumonia and, rarely, pleurisy; hyperaemia of the brain and its membranes, and also of the abdominal viscera, such as the spleen, liver, kidneys and intestines; and haemorrhages under the pleural, pericardial or cerebral membranes, while Henoch reports /

Morbid
anatomy

reports the occurrence of gastritis haemorrhagica in one of his cases. Although one or more of these gross lesions may have been found in different instances, the degree of severity being variable, there is none which occurs as a constant factor in the disease, and they are therefore to be considered as possible complications or secondary developments rather than as conditions which have any essential or causal relationship to the main condition.

Morbid
anatomy

The characteristic changes to be found in a well marked case are those in the skin and the tissues underlying it. The results of Parrot's examinations, as quoted in brief summaries by Henoch and by Crocker are as follows:- Extreme atrophy with consolidation of the skin, including the rete Malpighii the cells of which are scarcely visible and form a compact mass with ill-defined contours. The horny layer is unchanged and only looks thicker by contrast with the thinned rete and corium. In the subcutaneous fat, the fibres of connective tissue are more numerous than usual and thicker, and the fat itself is considerably diminished; the fat cells are smaller and their nuclei can be distinctly seen. Most of the fat cells are, /

Histology

are, as in every form of atrophy, almost or entirely deprived of their fat; they are shrivelled into an oval shape and have a great resemblance to the epidermic cells of the rete Malpighii. The blood vessels - especially those of the papillae of the skin are narrowed to such an extent that one cannot distinguish their lumen. Henoeh adds that in certain cases observed in his wards, dissection of the skin had yielded similar results, - drying up of the skin, with consolidation of its layers and atrophy of its tissue.

Histology

Ballantyne (B.M.J. 1890) in a report of one examination, says that section of the skin and subcutaneous tissue gave the sensation of cutting bacon rind, and that the subcutaneous cellular tissue had a peculiar white glistening aspect, quite unlike the yellowish appearance of the subcutaneous fat in a normal infant. There was no serous fluid to be expressed and no microscopic appearance of congestion. On microscopic examination, the outstanding feature of the sections was the presence of abundant brightly stained connective tissue, the fibres of which, increased in number and abnormally thickened, subdivided the adipose tissue into patches of varying size.

Ballantyne

The /

Ballantyne
on histology

The cells composing the connective tissue could be clearly differentiated, and there were also to be seen blood vessels relatively large as compared with those in the papillae, each surrounded with leucocytes, and projecting into the groups of fat cells. In the adipose tissue, it was seen that none of the cells had their normal amount of oil and some of them were devoid of fat; the nucleus in all was clearly visible, and in many a ring of protoplasm underlay the cell-wall. The papillae were not well marked and the cells of the rete Malpighii were ill-defined. The horny layer appeared to be normal. Ballantyne while recognising the general correspondence of Parrot's description with his own, considers that the lesion is something more than cutaneous consolidation with atrophy of the tissue. In his opinion, the primary change is in the penetration of a capillary into the fat cells, the capillary being surrounded or accompanied by leucocytes: from this there arises an increased formation of fibrous tissue and subdivision and atrophy of the fat cells.

Mensi, as I have already quoted, described the characteristics of one of his types to be atrophy of the skin with absence of the granular layer, the cells of /

of the cutis are closely packed together, and there is dilatation of the vessels with haemorrhages into the skin. In the other type he describes atrophy of the skin, with abnormal development of fibrous tissue round the fat. He holds that the main feature of sclerema is in the alteration of the cutaneous fibrous tissue.

Histology

Mensi

Finkelstein says that in this disease there may be hyperaemia of the muscles, with a small-celled infiltration alongside of each capillary.

Finkelstein

In the report of a typical case given by Waterhouse (Lancet 1906) there is a remarkable agreement with Ballantyne's description. He speaks of the abnormal resistance to the knife on cutting, the alteration of the panniculus adiposus from the appearance of loose yellow greasy fat to that of a dead white layer, soft but compact, resembling dried sebum and closely adherent to the skin. He also notes the remarkable absence of normal fat even in the omentum and round the kidney. There was abnormal density of the connective tissue of the cutis vera, with fibres extending into the subcutaneous tissue, and the blood vessels of the cutis vera had thickened walls.

Waterhouse

It is to be recognised that the microscopic conditions /

conditions found by different observers shew some degree of variance. I would suggest that these variations may depend on the individual conditions of the cases described. In a case characterised by very severe symptoms passing rapidly into death, it might be that the end would come before the increase of the fibrous tissue had occurred. For example, in the microphotographs shewn from Dr Findlay's case, there is no appearance of abnormal fibrosis, but it is to be remembered that the child died within twenty hours of the signs of sclerema being first seen by the mother.

If the symptoms were less violent, and the termination more gradual, time would be given for the development of fibrous tissue and for the atrophic changes in the adipose tissue.

From the various descriptions, one may conclude that the primary change consists in the infiltration with small cells of the true skin and subdermal layer, and, possibly, of the rete Malpighii. As a secondary change there would occur an increase in number and density of the connective tissue fibres, with alteration in the shape and contents of the fat-cells, and these latter features, when present, would /

would be the most prominent microscopic features.

Aetiology.

Aetiology

To give some rational explanation of the cause of these morbid phenomena, many theories have been propounded, ranging from the doctrine of maternal impression set forth by Usembezius to that of septic intoxication suggested by Eustace Smith in 1898. Some of the earlier observers considered that the lesions were akin to erysipelas, or due to some inflammatory process, while others saw an analogy to phlegmasia alba dolens. Parrot, as has been mentioned earlier, considered that the induration was one of the features of athrepsia, due to faulty nutrition, over-crowding and the like, and in this he was in agreement with Underwood's description of it as an hospital disease, at a time when the unsatisfactory state of the hospitals did much to cause disease. But debility arising from the unfavourable circumstances which are often associated with sclerema, is not in itself sufficient to cause this peculiar affection. This is proved by the fact that the disease may occur in an infant otherwise healthy and free from the unhygienic influences of syphilis, poverty, immaturity or squalor. Further, if these unfortunate conditions which are so commonly encountered, or if any group of them /

them were the immediate cause of the ailment, rather than predisposing influences, then sclerema would be almost certainly a disease of common occurrence instead of being one of the rarities of medical practice.

It was suggested by Léger in 1823 that one of the main factors was the effect of cold in causing coagulation in the subcutaneous tissues of the new born. Many years later, in 1881, Langer held a similar view, that the condition was essentially a physical change in the adipose tissue due to cold - an actual freezing or solidification of the fat, - and this he based on his demonstration of a marked difference between the fat of infants and that of adults. This observation was confirmed by Knöpfelmacher, and it consisted in the fact that the adipose tissue has in infancy more stearin and palmitin and less olein than in adult life. As a result of this excess of fatty acids, the difference being as 31 per cent to 10 per cent, the fat of the new born solidifies at 89.6° Fahr. while that of adults solidifies below 32° Fahr. This theory was accepted by Northrup (Archives of Pediatrics 1890) and it has received a certain amount of support but, to my mind, it is untenable. It is not consistent with the fact that the induration /

Theory of
Hypothermia

induration generally begins in the lumbar and gluteal regions, which are usually protected from cold, rather than beginning in exposed parts such as the face and extremities. It is not a probable cause of an induration which is found in irregular patches instead of being generalised over an area which might have been exposed to cold. It does not account for congenital cases. It does not explain the microscopic findings of increase of the fibrous tissue with actual atrophy of the fat cells, and the loss of some or all of their fat. Still further, if the essential lesion were a congelation of the adipose tissue, the continuation of the condition would be incompatible with the occurrence of pyrexia. In the case recorded by Waterhouse (Lancet 1906) there were variable degrees of pyrexia, reaching as high as 105.6^o Fahr. but without any apparent difference in the indurated areas. For these reasons principally, I hold that the contention is unsound which would ascribe the cause of the disease solely to the results produced by cold on the adipose tissue.

Certain other suggestions, by which the onset of the disease has been respectively attributed to patency of the foramen ovale or other disturbances of the circulatory system, to pulmonary lesions, to shortness of the intestine or other gastro-intestinal or hepatic affections, /

affections, do not seem to call for much consideration, in view of the fact that typical cases of sclerema may occur without the coincidence of any such conditions. Ballantyne, in his 'Diseases of the Foetus' devotes attention to various points as possible aetiological factors. He refers to the incidence of sex of the infants affected, to the exact date of the first symptoms, to the state of health of the infant or of one or both parents, to the diet of the child and the season of the year when the disease is most likely to be found. It would serve no useful purpose to give a mere transcription of his statements, to which reference is easy, but his conclusions are that none of them has any direct bearing on the causation of the disease, and that, as predisposing causes, they are only of importance in so far as they contribute to the lowering of the vitality. He indicates his general agreement with the opinion that the primary cause is to be found in some trophic disturbance of the nervous system, and it is to be noted that Coats draws attention to the similarity of the lesion in sclerema with those found in certain tropho-neuroses. This theory of nervous influence had been put forward first by Liberali in 1818, and, with various modifications, had been adopted by different writers, including Angel Money /

Theory of
Tropho-neurosis

Money (1889) and G. Somma (1892). The opinion of the last named writer is quoted at some length by Ballantyne, and it may be summarised as follows. Somma accepts as proven the existence of thermogenetic, thermolytic and thermotactic centres in the brain; from this he concludes that, given a congenital debility of the infant as a predisposing cause, there might follow chilling of the surface as a determining cause, and that from this centripetal stimulation of the thermotactic centres, there would follow the efferent impulses resulting in the local changes in the subcutaneous tissues. Various objections may be raised to the acceptance of this or any allied theory of causation. First, the existence of heat-regulating centres in the brain is chiefly hypothetical. Secondly, no cerebral lesions have been found as constant factors. Thirdly, congenital debility is not a necessary antecedent; it has been shewn that the disease may occur in a child apparently in good health. Fourthly, chilling of the cutaneous surface is not an invariable concomitant; this is demonstrated by the facts that sclerema may occur in warm weather and, still further, that the disease may be actually congenital, its manifestation having begun in utero, without /

Theory of
Tropho-neurosis

G. Somma

without any exposure to cold. Fifthly, while the trophoneurotic theory might reasonably give some explanation of the subcutaneous lesions, it does not so readily account for jaundice, subserous haemorrhages or broncho-pneumonia which are commonly found as complications.

The effects of internal glandular secretions have also been taken into account as possible causes of the disease, and reference may be made to the somewhat comparable condition found in myxoedema, where also there is an induration which does not pit on pressure. This aspect has been dealt with by Mensi (op.cit.) who concludes that there is no reason to ascribe sclerema to any change of the internal secretions. He suggests that possibly some variation of the thyroid secretion, either by increase or diminution of its efficiency, might act as a predisposing cause and might also assist to generate the toxic agent. It may here be stated that in the case which was under my care extract of thymus gland was used according to Carpenter's suggestion, but it seemed to produce bad results and after three weeks it was discontinued.

Internal
secretions

In the light of modern pathology, one is forced to consider the question of microbic effect in the production of such lesions as we find in sclerema, and it /

it would seem that in this direction we are most likely to find a satisfactory explanation. This theory was put forward in this country by Eustace Smith in 1898, and it has been further elucidated by others, such as Schmidt, Jemma and Comba, whose results would serve to shew that there is no specific micro-organism but that probably various organisms either as a simple or mixed infection of streptococci, staphylococci or Friedländer's bacillus, may act as causal factors in the disease. A toxæmia of this kind would reasonably explain the multiplicity of conditions which have been described. It would account for the evidence during life of lowered vitality; for the pyrexia which is sometimes present or, by extreme prostration, for the subnormal temperature which may be one of the more striking signs of the disease; for the icterus mentioned by Hensch as a very common symptom, which, in the absence of any hepatic lesion may be of the ordinary type found in many healthy children, but which may also be due to some toxæmic destruction of the blood-corpuscles. Of the conditions found after death, septicaemia would also afford a rational solution. The microscopic findings of subcutaneous fibrosis, destruction of the fat and increased leucocytosis beside the capillary blood vessels are all suggestive of some toxic irritant. The other lesions which have been found /

found as common though not invariable, complications - bronchitis, bronchopneumonia, haemorrhages into the serous cavities, gastritis haemorrhagica quoted by Henoch and the "polyarthrititis rheumatica" found in a case recorded by Demme (1882) - cannot reasonably be explained as the results of anything but septicaemia. In a contribution by Money (Lancet 1888), he records the occurrence of paralytic symptoms in cases which may have been sclerema, and suggests the theory of a trophoneurosis. Acceptance of his observation, however, does not necessarily militate against the microbic theory. It is well known that evidences of nerve-injury may be found in some diseases which are undoubtedly of microbic origin. The post-diphtheritic paralysis due to the Klebs-Loeffler bacillus, the nervous phenomena due to the bacillus tetani, the general paralysis following the spiræchaete of syphilis, and, possibly, the symptoms of cases of epidemic poliomyelitis acuta, are all instances of this type, although in the last mentioned no specific organism has yet been isolated. Thus Money's observation of paralysis occurring in sclerema is not destructive of the microbic theory but, by analogy, would tend to strengthen it.

Prognosis.

The /

The prognosis in a case of sclerema must be always guarded, usually grave, but not invariably hopeless. It will in some measure depend on the extent of the local lesions and also on the presence or absence of complications. It will vary unfavourably or favourably according to the gravity of the signs exhibited, such as the record of the temperatures, the state of the heart or other indications of prostration, the lower degrees of temperature and the slower pulse giving the worse prognosis. It will be affected markedly by the general circumstances of the case, being better in the case of a mature child of sound constitution born into favourable surroundings than in that of a premature and weakly infant in adverse social and climatic conditions. It will also depend on the infant's personal degree of resistance to, or immunity from the actual materies morbi; in this respect, it is possible that hereditary tendency may have some bearing on the prognosis, as in the record by Money (Lancet 1888) of three successive cases in one family which were possible cases of sclerema accompanied by paralytic symptoms.

Treatment.

In dealing with a disease where there is lack of full knowledge as to the cause, the first direction of the therapeutic measures must be towards the treatment of /

Prognosis

Treatment

of symptoms. In sclerema the wide range of theories as to its cause and its essential features has led to a great variety of means of treatment, many of which have fallen into disuse. The necessity is obvious to place the child in the most favourable hygienic surroundings and to maintain its strength, as far as possible, by the administration of suitable nourishment. In my own case, an extemporised incubator was used with satisfactory results. An ordinary clothes-basket was taken, and pigeon-holes were cut in its lower segment, each one large enough to admit a lemonade bottle full of hot water. Over the layer of bottles a bed was made, and the inside of the basket was lined so that the child was almost completely covered in. Each bottle in turn was slipped out, refilled with hot water and reinserted, so that the temperature was maintained at a fairly constant level. More elaborate methods to obtain the same result have been described by different writers.

As a remedial measure many have used warm baths from 90° to 99° Fahr, with or without the addition of mustard or aromatic substances, the duration of each bath being from fifteen to twenty minutes. Underwood and some others recommended the use of vapour baths, while others again applied warm lotions to the skin.

Friction /

Friction of the body and limbs has been considered serviceable, and also massage, with theunction of olive oil, camphorated oil or mercurial ointment, apparently with benefit. In my case I prescribed gentle rubbing with Unguentum Iodex (Menley & James, London), a composition having free Iodine (5%) with an oleaginous base. This preparation has the advantage that, even with repeated applications, it does not cause hardening or vesication of the skin, and it was used daily for about four months. I am of opinion that it was beneficial, possibly from the alterative effect of the Iodine, possibly by its promoting absorption of the induration, possibly too, from the well-known germicidal action of Iodine.

In the matter of internal medication, we find an extensive list of preparation which have been used and recommended. Diaphoretics, diuretics, emetics, stimulants and aperients have all had their advocates, while digitalis, mercury and cinchona have also had their supporters.

Some have used drugs capable of subcutaneous administration, because of the difficulty of swallowing, and the inhalation of oxygen has also been recommended. In the case which was under my care, the first indications were to combat the cardiac weakness. Whisky, in /

in small quantities, was given by the mouth: 'Digalen', Treatment
 prepared by Hoffmann, La Roche & Co., was used hypoder-
 mically, and on two occasions, a subcutaneous injection
 of Adrenalin Chloride (soln. 1 - 1000) was also given.

For the general condition, extract of thymus
 gland, as suggested by Carpenter, was used for about
 four weeks. Of the 'tabloid' preparation of thymus
 gland, one grain daily was given at first, then one
 grain twice daily, and latterly two grains twice daily.
 After about three weeks the mother noticed that the
 child became distinctly pallid for a while after each
 dose, and its use was therefore stopped.

If it should happen for me again to have a case
 of sclerema under my care, I would think it right to
 make a bacteriological examination of the blood, with
 the hope of discovering one or more causal organisms.
 Pending any such information and the possible prepara-
 tion of an autogenous vaccine, I would think it reason-
 able to try the administration of a polyvalent vaccine,
 and to judge by the results as to the desirability of
 its repetition.

General conclusions.

On general consideration of this strange disease,
 one is forced, by the limited opportunities for
 observation, to draw conclusions from the cases re-
 corded /

Conclusions

recorded by others, rather than from a wealth of personal experience. The main points which I have endeavoured to establish as being in accordance with our present knowledge are the following:-

- (1) Sclerema neonatorum is to be recognised as a definite disease which may be found with or without the association of other recognisable morbid conditions.
- (2) It may vary considerably in degree as to the extent and severity of its manifestations, and also as to the gravity of the progress.
- (3) The name properly includes the so-called 'pseudo-sclerema', the differences between true and false sclerema being only those of degree.
- (4) While the disease is a rare one, it is probably of more common occurrence than the usually accepted ideas would suggest.
- (5) It is found at or shortly after birth, and usually in circumstances which are unfavourable to the welfare to the infant.
- (6) The most characteristic feature is in the palpable subcutaneous induration, with which there may be associated abnormalities of temperature, most notably in the direction of its being subnormal.
- (7) Visceral /

- (7) Visceral changes may occur as complications, in the form of inflammations, haemorrhages or serous exudations.
- (8) The description of an 'oedematous' type of sclerema is probably unwarranted, the oedema in such cases being a complication rather than an essential feature.
- (9) Microscopic examination of a well marked case would shew subcutaneous infiltration with small cells, and increased fibrosis, with atrophy of the adipose tissue.
- (10) The origin of the disease is probably to be found in some microbic infection.
- (11) Treatment must be largely symptomatic, but also some form of treatment should be sought which would counteract the toxaemia.

PUBLICATIONS TO WHICH REFERENCE HAS BEEN MADE BY ME

FOR THIS THESIS.

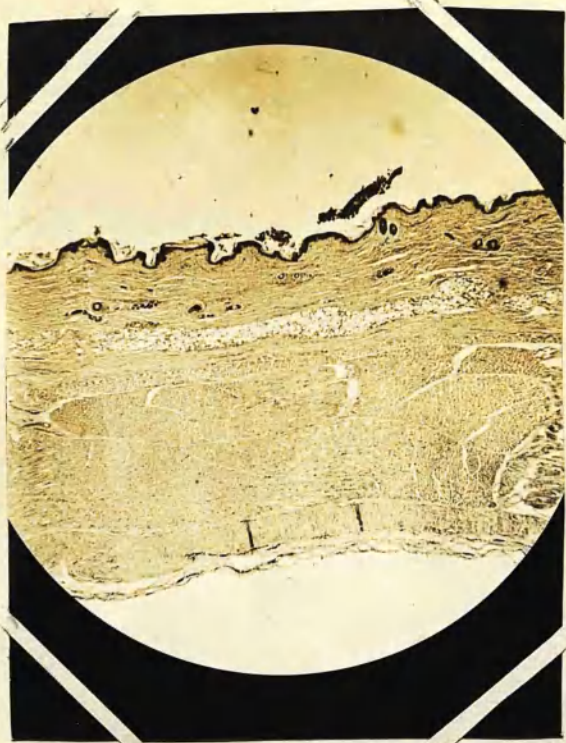
Abraham	Allbutt's "System of Medicine"	1901
Ballantyne	British Medical Journal	1890
Ballantyne	"Diseases of the Foetus"	1895
Barrs	British Medical Journal	1889
Bouchut	"Diseases of Children" (trans. by Bird)	1855
Bristowe	"Theory and Practice of Medicine"	1890
Carpenter	Lancet	1906
Coats	"Manual of Pathology"	1889
Duhring	"Diseases of the Skin"	
Evanson & Maunsell	"Diseases of Children"	1847
Finkelstein	"Lehrbuch der Säuglingskrankheiten"	1908
Garrod	"Encyclopaedia Medica"	1902
Goodhart	"Diseases of Children"	1899
Gould & Pyle	"Cyclopaedia of Medicine and Surgery"	1900
Henoch	"Lectures on Children's Diseases" (Sydenham Socy.)	1889
Jacobé	"Therapeutics of Infancy and Childhood"	1897
Kennedy	"Management of Children"	1825
Langmead	"Diseases of the Newborn (Cautley)"	1913
Mackenzie	British Medical Journal	1889
Mensi	"Zeitschrift für Kinderheilkunde"	1911
Money	"Disease in Children"	1887
Money	Lancet	1888, 1889
Monro	"Manual of Medicine"	1911

Publications
Particulars to which reference has been made by me
for this Thesis. (Contd.)

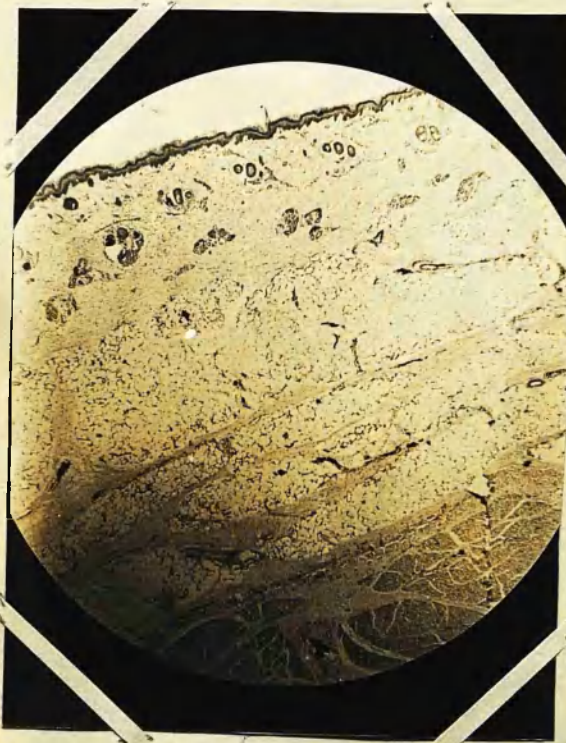
Osler	"Practice of Medicine"	1909
Parrot	"Athrepsie"	1877
Robinson	"Manual of Dermatology"	1885
Rommel	"Diseases of Children"	
	(Pfaundler & Schlossmann; trans. by Shaw & La Fétra)	1908
Shepherd	"Diseases of the Skin" (Bangs & Hardaway)	1898
Simpson	"Obstetric Memoirs"	1855
Taylor & Wells	"Diseases of Children"	1901
Walker	"Introduction to Dermatology"	1902
Waterhouse	Lancet	1906



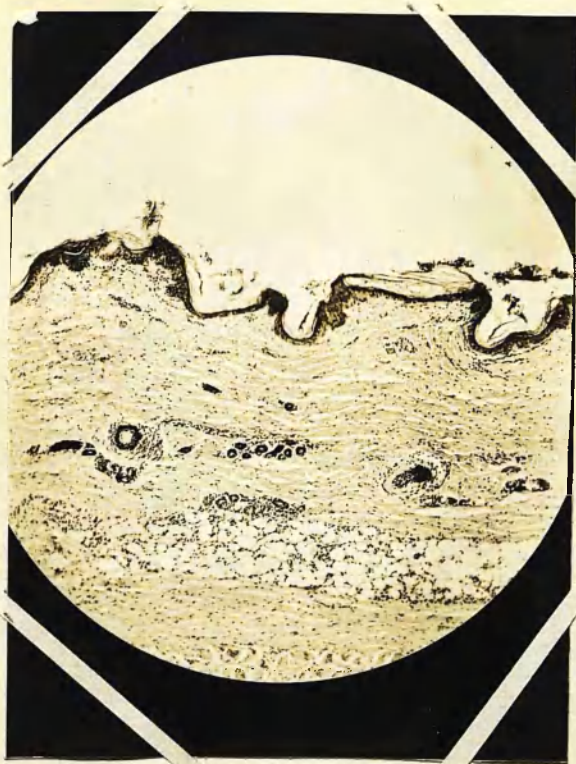
Photograph of Dr. Findlay's case, aged 13 days.
Congenital Syphilis; Pemphigus; Sclerema neonatorum.



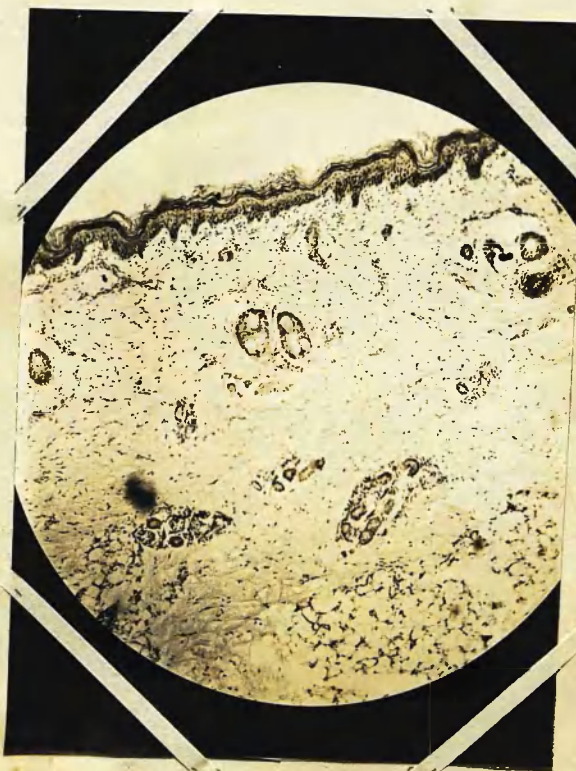
Normal skin from
abdomen of infant
of 3 months. X 24



Skin from thigh
of Dr Findlay's case
of Sclerema, 14 days
X 24



Normal skin from
abdomen of infant
of 3 months. X 68
(streaky appearance
due to artefacts from
the microtome.)



Skin from thigh
of Dr Findlay's case
of Sclerema, 14 days
X 68



Photograph of case described by W. S. Paterson.
Aged 19 months (May 1914); weight $19\frac{1}{2}$ lbs.