REFRESHER COURSE FOR GENERAL PRACTITIONERS

PINK DISEASE

BY

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Pink disease is a clinical entity met with only in children. Most cases occur between the ages of 6 months and 3 years. The disease shows no predilection for either sex. Breast-fed infants enjoy no special immunity. There is a marked seasonal incidence, the majority of cases in this country occurring between December and April. Although statistics are not available there are grounds for believing that year-to-year variations in the incidence of pink disease are related to the severity of the winter, and that children in rural areas are more liable to develop the condition than those living in cities.

The condition is of particular importance to the family doctor because in the early days it may give rise to suspicions of one of the commoner infectious diseases, and because the exacting demands of the patient over a prolonged period inevitably disturb household routine. Treatment is dependent for success upon patience on the part of both family practitioner and parents, and upon the family doctor's ability to retain the confidence and co-operation of the mother.

The Fully Developed Clinical Picture

In its fully developed form the clinical picture of pink disease is unmistakable. The appearance of the child is one of abject misery. He is peevish and fretful, and whines unceasingly. Pathetically resigned to the misery of his own condition, he vents his irritability on others only when disturbed. He shows no response to any form of attention and is completely disconsolate. He resents approach, but is equally discontent when left to his own resources. All interest in toys is lost. Food has no attraction, no

FIG. 1.—A typical attitude: the child is scratching his back, chewing his arm, and rubbing his feet together.

matter how varied or how attractively prepared. The child dislikes facing the light. Photophobia is common but may be absent, and when present it may be severe or slight. Aversion to light leads the child to turn his back to the window and to bury his head in the pillow or assume the so-called knee-elbow position. Whatever his position, he is never still, continually scratching himself, rubbing his hands and feet, or chewing his fingers (Fig. 1). If weakness keeps him recumbent he wriggles and writhes incessantly (Fig. 2). He may even pull his hair out. In its extreme form restlessness ceases only when, with fatigue added to weakness, the child falls asleep through sheer exhaustion. When this takes place sleep usually occurs

during the day, the child continuing to be noisy and restless during the night.

The erythematous changes which have given rise to the name pink disease are to be looked for in the hands and feet. When dry and warm the extremities are bright pink in colour. If cold and clammy they have a bluish-red

FIG. 2.—The child wriggles and writhes on his abdomen. There is an extensive erythema, wasting is severe, and the hair has fallen out.

appearance. The discoloration is most pronounced in the tips of the fingers and toes. Proximally it lessens in intensity and is scarcely perceptible above the level of the wrists and ankles. The erythema may vary from time to time in intensity and, less often, in extent. It may disappear completely, only to reappear. In addition to being discoloured the extremities are "puffy" or swollen, but there is no pitting oedema.

The skin over the affected parts has a glazed appearance and is often the site of local desquamation. Extremities showing these changes have been aptly described as looking like "raw beef" or "washer-woman's hands" (Fig. 4). Sensory changes occur and account for the local irritation evidenced by infants and for the feelings of tingling and numbness described by older children. Handling or manipulation of the extremities does not cause pain. The erythematous changes are not always confined to the hands and feet. Bright redness of the cheeks or of the nose may first attract attention, but is not met with in the

FIG. 3.—Another typical attitude in a child who has been disturbed. Peevishness, photophobia, wasting, hypotonia, and swelling of the feet are all evident. (Professor Charles McNeil's case.)

FIG. 4.—Showing the picture of abject misery, the “raw beef” hands, and pronounced photophobia. (Professor Charles McNeil's case.)

absence of changes in the hands or feet. Excessive sweating is common and may become so profuse that the patient is literally drenched in perspiration many times daily. Persistent and profuse sweating in an afebrile child of under 3 years of age should always raise the question of pink disease. Sweating is accompanied by thirst, but by a thirst which is not commensurate with the amount of fluid lost via the skin; the former gives rise to a sudaminal rash which adds to the patient's acute discomfort. Occasionally sweating is absent. In that event the dry powdery skin covering the emaciated trunk and limbs contrasts strikingly with the sodden condition of the hands and feet.

Invariably lack of sleep, loss of appetite, unrelieved fatigue, and profuse sweating contribute to a severe loss in weight and to a state of unmistakable physical weakness. The abdomen may be scaphoid. These developments are accompanied by a generalized loss of muscle tone. The degree of hypotonia is so extreme as not to be entirely accounted for by the physical condition as a whole. Individual muscles or muscle groups lack bulk—and such bulk as they possess moulds readily in response to the gentle pressure of the bed-clothes with which they are in contact. Equally striking is the impression of a jelly-like consistence obtained by the examiner on gently palpating or handling the muscles. The tendon reflexes may be diminished or absent, but there is no muscular paralysis in the strict sense of the word.

Tachycardia is present in all cases, and is usually so pronounced that it can be confidently recognized despite the fretful reaction of these children to examination. The blood pressure is appreciably raised, but confirmation of this is not necessary for the purposes of diagnosis. In an uncomplicated case the temperature is normal or slightly below normal: and there are no characteristic changes in the blood picture.

Development of the Picture

Such, then, is the clinical picture in fully developed form reached gradually over a period of several weeks. Rarely is the onset sudden. Usually it is gradual. In retrospect the earliest signs or symptoms are recognized as having been those of a mild upper respiratory catarrh. The catarrh may consist of little more than a discharging nose. There may or may not be slight fever. Not infrequently this is the only febrile phase during the entire course of an uncomplicated case of pink disease. Sometimes the catarrh tends to persist. More usually it simulates a mild coryza, disappears in less than a week, and is regarded at the time as having been a passing incident. A few days elapse, and then it is noticed that the child is not his usual self. The mother is disappointed that he has not improved according

to her expectations and may even decide that “perhaps after all he has had an attack of influenza.” More than anything else the child remains strangely restless: peevishness is added to restlessness, appetite is lost, and sleeplessness follows. In these circumstances any suggestion of photophobia should lead to a tentative diagnosis of pink disease even in the absence of discoloured or swollen extremities.

The more gross changes which occur in the hands and feet are preceded by the appearance of a general erythema covering limbs and trunk alike. The erythema is of a transient nature. After a varying interval of time it loses its general character and fades away over the trunk and limbs while becoming intensified in the hands and feet. When this stage has been reached there can be no mistaking the diagnosis. Symptoms may continue to worsen, but not necessarily so. Improvement can, and often does, set in without development of the complete clinical picture which has been described. In a case of average severity the clinical climax is reached in some six to eight weeks.

The Stage of Recovery

Whether signs and symptoms are of severe or moderate degree the disease runs a long course. In the mildest cases recovery takes six to eight weeks (Fig. 5). In general it

FIG. 5.—A child has passed the clinical climax but still shields his eyes from the light. Note the loose “pendulous” musculature of the limbs.

can be said that the time taken to reach the stage of clinical climax corresponds approximately to the further time required for symptoms to disappear completely. In the severest cases twelve months may elapse from the onset of the disease before symptoms finally and completely disappear. The trials of slow recovery are aggravated by the tendency for progress to be erratic and for relapses to occur.

Complications

Estimates concerning the incidence of fatal results vary from 5 to 10%. Intercurrent infections constitute the main threat to life. Of these the most serious is broncho-

FIG. 6.—Secondarily infected skin lesions. Wasting is severe and the extremities are swollen and discoloured.

FIG. 7.—Swollen, deeply coloured, unhealthy gums. The incisor teeth subsequently fell out.

pneumonia. Exceptionally, sudden unexpected death follows acute cardiac failure. Pyelitis and otitis media may develop and the stools may become fetid. Trophic lesions are common, more especially in severely debilitated cases running a prolonged course. Rashes and other skin lesions which have become secondarily infected are slow to heal (Fig. 6).

If there is secondary infection the glands draining the affected areas become enlarged and sometimes tender. Fingers which have been rubbed or chewed are apt to become the site of resistant pulp or nail-fold infections. The hair may fall out. Frequently the gums are unhealthy in appearance, being swollen and of an unusual deep mulberry colour (Fig. 7). Gingivitis and stomatitis soon follow if measures are not taken to prevent the child biting his hands and fingers. When this occurs the teeth may fall out. Rectal prolapse is the commonest of the less serious complications. Children recovering from pink disease do so completely, except in so far as chest complications may result in varying degrees of pulmonary collapse and septic skin conditions may leave permanent scarring. Second attacks of pink disease do not occur.

Aetiology

The cause of pink disease is unknown. Allergy, endocrine dysfunction, toxicity due to arsenic and ergot, and photo-sensitivity have all been suspected in their day. Within recent years it has been suggested that mercury contained in medicinal tablets, ointments, or teething powders may be a contributory cause. This view is based on the finding of large amounts of mercury in the urine of children in whom a diagnosis of pink disease has been made and who have been given mercury in one of the forms mentioned. By no means all children excreting mercury in the urine show signs of pink disease, and it is possible that the development of the condition may be dependent in part upon individual susceptibility to mercury intoxication.

The significance of mercury as an aetiological factor must be looked upon as *sub judice*. The same may be said concerning prevailing theories that pink disease is a deficiency disease or a virus infection. Suspicions that pink disease was a form of polyneuritis stimulated trial of intensive vitamin B and liver therapy. Pioneer work along

these lines gave promising results which have not been universally substantiated by subsequent clinical experience or by experimental studies. Those who favour the theory that pink disease is a virus infection are influenced by the catarrhal symptoms at the time of onset; by the liability of cases to occur in small local "crops"; and by recorded findings, at the few necropsies which have been carried out, of diffuse infiltrative lesions involving the cord and brain substance and of degenerative changes affecting the peripheral nerves.

Management and Care

The diagnosis of pink disease calls for a frank explanation of the implications. There is no specific treatment. Parents are readier to accept what is, after all, an unpalatable fact if they are at the same time told of prevailing doubts concerning aetiology. Mention of infection will prompt the question, "Is it catching?" The parents can be unhesitatingly reassured that there is little likelihood of any other member of the household contracting the condition. Recorded instances of two children in the same household suffering simultaneously from pink disease are few in number. On the other hand, it is not altogether rare for two members of a family to develop the condition at widely separated intervals—usually of years.

Inevitably the mother or father will ask concerning the desirability of hospital care. Many factors have to be taken into account in giving an answer. Whatever the ultimate decision the parents must know of their child's increased susceptibility to secondary infections and of the risks of his contracting such an infection in hospital. The mother should be told, and told with conviction, that she has it in her power to make a special contribution to the care of her child.

No matter the ideal, however, there is no escaping the practical difficulties. The mother may be too exhausted or her duties in the home may be such as to rule out any question of the child's being retained at home. Other circumstances peculiar to the home may point to the necessity for transfer of the child to hospital. So may the condition of the patient himself. The state of his skin and mucous membranes may call for more continuous nursing care than the district nurse can be expected to give; or the child may be so reluctant to take food that gavage in hospital is essential. In giving his advice the family doctor will be influenced by his knowledge of the facilities offered by whatever hospital he has in mind.

If it is decided to treat the child at home, full advantage should be taken of any home nursing service in the locality. Provided always that the suggestion appeals to her, steps should be taken to relieve the mother of some of her routine household duties by applying to the medical officer of health for the services of a home help. Alternatively, the help of an understanding relative or neighbour can be enlisted. Where arrangements of this sort are impracticable, a not altogether satisfactory compromise can sometimes be effected by temporarily admitting the child to hospital during such time as the mother gets a much-needed short rest.

For home treatment a room should, if possible, be selected which is quiet and well ventilated, which can be shaded without being completely darkened, and in which there is space for a couch or bed where the mother can rest during such time as the patient is asleep. An even room temperature in the neighbourhood of 60° F. (15.5° C.) should be maintained. Unless compelled by weakness to remain in bed the child should be allowed up for short periods once or twice daily, provided that he does not over-fatigue himself and provided that he is protected from

draughts. With the disappearance of tachycardia the patient can be allowed out of doors, but must in the first instance be protected from direct sunlight, extremes of temperature, and boisterous winds.

The child should be given a warm as distinct from a hot bath morning and evening. In order to soothe and lessen skin irritation boric crystals (2 oz.—57 g.) should be added to each gallon (4.5 litres) of bath water. A preferable alternative is to bath the child in warm normal saline. Soap should not be used. Bathing must be followed by careful drying with a warm towel and by the application of pulverized fuller's earth or of a high-grade *baby* dusting powder. Calamine lotion is commonly used, but is less effective. Special attention should be paid to situations where there are opposed skin surfaces. The eyes should be bathed and the mouth rinsed with normal saline or boric lotion. Clothing must be light, and the garments next to the skin should be of cotton or silk. Frequent changes of clothing are necessary to avoid chilling while sweating persists. Cardboard splints may have to be applied to the limbs to prevent rubbing. Cotton mittens and socks are useful in protecting the hands and feet.

Feeding

Every effort must be made to ensure that the child obtains adequate nourishment despite his lack of appetite. If the mouth and gums are tender he should (depending upon his age) be fed by feeding-bottle or by a feeder to the spout of which a soft rubber tube has been attached. The use of metal spoons should be avoided as being likely to cause pain. Nourishment should be given frequently—hourly if necessary—in small amounts, and lukewarm or cold according to the child's preference. According to age, chicken jelly, coddled egg, barley jelly, fruit jelly, the cream of fresh milk, and ice-cream (from a reliable source) can be offered. With improvement of appetite in a child of over 9 months choice can be extended to include "smooth" items of semi-solid nourishment such as Benger's food, cornflour pudding, carefully prepared oat flour, egg custard, and fruit juices. At a later stage calves' liver, chicken, and rabbit (all of which should be pounded) and creamed vegetables can be gradually added. Hard or "crumbly" items of diet should not be given until the child shows a readiness to take them. At all times fluids should be allowed generously in the form of diluted milk, provided it does not prove nauseating, or of fresh fruit juices, suitably diluted and sweetened. The aim should be to establish the child on a full diet as soon as he is able to tolerate it.

Although there is no proof that pink disease is of itself a deficiency state the debilitated condition of the child with pink disease requires that an adequate vitamin intake should be assured. In a severe case it is a wise precaution daily to give cod-liver oil (in amounts corresponding to 1,500–2,000 i.u. of vitamin D), ascorbic acid (50 mg.), and one of the reliable preparations of the vitamin B complex.

Sedation

The restlessness and lack of sleep call for sedation. Chloral hydrate is the drug of choice (1–3 gr.—0.06–0.2 g.). On occasion potassium bromide (2–5 gr.—0.13–0.32 g.) is effective when chloral fails, but is apt to give rise to a bromide rash. Sometimes a combination of the two drugs gives the best results. Exceptionally, recourse to phenobarbitone for a limited period of time is necessary. Medicaments, no matter what their form, known to contain mercury should not be given; it should be remembered that teething powders are suspect in this connexion. The risk of secondary infections has been mentioned. Penicillin or

sulphonamide therapy is indicated when these threaten, but is not to be regarded as an essential part of routine treatment. Atropine has been employed as a means of controlling the sweating, but the advantages are more than offset by the aggravation of thirst which results. Artificial sunlight and massage therapy are of value after symptoms of irritation and photophobia have disappeared and when the stage of convalescence is being approached. A simple bitter tonic is useful at this stage.

Ideally, the city child should be sent to the country for convalescence—preferably inland rather than to the seaside. In her own interest the mother should if possible accompany the child. Follow-up care must have as one aim the prevention of delayed respiratory complications.

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DISORDERED SOCIETY

BY

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Working in an international organization for the care of refugees, in Austria, over a period of some 18 months, my attention has been drawn to interesting behaviour patterns, which, as I shall try to show later, seem to lend themselves to a more general application—that of human behaviour in the insecure and disordered world that we live in at present. After all, the refugee is the physical example of the insecurity that may overtake any of us.

The subject being complicated and controversial, only brief mention can be made of those working for or with the refugees and their motives for doing this work, and of the attitudes of the members of the occupying Power, the Austrian authorities, the voluntary agencies (mostly of various religious denominations), and members of missions sent by Governments to select candidates to emigrate to their various countries.

In the area to which I refer the organization employed an international staff of 45, comprising 12 different nationalities, to supervise the care of the refugees. A temporary organization of this type and size had not existed before, and many of the posts had no parallel elsewhere, so that the holders often had had entirely different training for doing the same job. For instance, "eligibility officers" decided whether a refugee should be accepted as a responsibility of the organization (with enormous advantages to the small proportion of stateless people who were entitled to this privilege), and "resettlement officers" gave the opportunity for and facilitated emigration. Such professions have not existed before and probably have no future to them, yet their incumbents were of overwhelming importance to the refugees.

The members of the occupying Power mostly dreamt of returning home, and anticipated fate by living a life in these alien fields as akin to that at home as possible. They were nominally all-powerful in the life of the organization and of the refugee, but—with rare, though important, exceptions—avoided contact with the problem except at the highest levels, where it could not be avoided. At times the Austrians performed their obligations to the refugees, which were considerable, only under duress.

The mission members were treated with particular respect, because it was they alone who could achieve the most