

Three weeks after admission the possibility of actinomycosis was first considered, and this led to biopsy of the tumour a week later (20.7.53), which confirmed the diagnosis (sulphur granules in the fluid; *Actinomyces bovis* on culture).

Sulphamezathine in dosage of 2.0 grammes/day was now introduced, to supplement the penicillin; also potassium iodide in increasing doses. On 24.7.53 he looked ill; within three days the temperature had reached 104° F., by which time the appearance of a generalized erythematous rash suggested a diagnosis of drug intoxication. All medicaments were stopped, and within thirty-six hours the temperature was normal again.

Administration of streptomycin was commenced in dosage of 600 mg. b.d. Iodide was next reintroduced, without apparent ill-effect, and increased to a dose of 10 grains t.i.d.

On 6.8.53 a small lump appeared at one end of the biopsy wound, suggesting imminent breakdown (Fig. 1); this was averted, however, by 6 exposures to deep X-ray, given on alternate days.

At the time of presenting the case, three months' treatment with streptomycin was completed. Clinically, progress was satisfactory; he was up and about, afebrile, and gaining weight. A recent blood sedimentation rate was normal and a radiograph of the chest showed considerable improvement in the changes previously described. The original swelling had subsided, but there were two small fluctuant areas in the line of the biopsy incision.

POSTSCRIPT (November 1953).—The two fluctuant areas were incised and curetted; sulphur granules found. Streptomycin and potassium iodide are continuing. N. J. R.

**Generalized Lipodystrophy. Infectious Mononucleosis. Mild Infantile Hemiplegia.**—JOHN DAVIS, M.B. (for J. P. M. TIZARD, M.R.C.P.).

S. C., female, aged 31 months, the older child of young, healthy non-consanguineous parents, was born at full term after an uncomplicated pregnancy and labour, weighing 7 lb. 5 oz. She was healthy, and developed normally as an infant, but never exhibited a very good appetite. She was noticed to drag her right leg when she learned to walk: a symptom which her parents related to an immunizing injection given into the right thigh a month or so previously.

Her present symptoms appeared when she was 20 months old, after a severe but otherwise typical, attack of measles: while convalescent she lost all appetite, and several pounds in weight, and was found by her doctor to have visibly enlarged lymph glands in all major nodes. She had a second undiagnosed febrile illness of a week's duration during her mother's second confinement in January, and following this, wasted more rapidly and became very fractious and miserable. She was admitted to Paddington Green Children's Hospital in March 1953, and found to exhibit the abnormalities reported below. Diagnoses of lipodystrophy and infectious mononucleosis were made at that time. Since her discharge, she has gained several pounds in weight and grown rapidly taller, but her physiognomy has not altered and she has continued to run a low fever (100° F. P.R. nightly). She is said to drink and void excessive quantities of fluid, and continues to wet her bed almost every night. Her hair which had previously been quite straight, became curly in August.

*Physical examination.*—A tall, muscular-looking girl, with prominent veins and no appreciable subcutaneous fat anywhere. There is a suggestion of exophthalmos, and she is alert and nervous (Figs. 1 and 2).

T. 100° F. P. 120.

Height 3 ft. 2½ in. Weight 2 st. 6 lb. 9 oz. (Ht. 90. Wt. 80—percentile).

C.V.S.: Moderate persistent tachycardia. R.S.: Trachea deviated to right. C.N.S.: Slight wasting of, and increase in tone and deep reflexes in the right leg as compared to the left.

Abdomen: Liver enlarged 1 fingerbreadth; spleen palpable.

E.N.T. healthy.

Skin: Folds of skin, pinched between finger and thumb equally meagre in all areas; face, trunk, arms, buttocks and legs all being affected (Fig. 2).

Glands: firm, discrete mobile, moderately enlarged glands palpable in occipital, cervical, axillary, epitrochlear, and inguinal nodes.

B.P. (after crying) 120/90.

*Biopsy of skin from buttocks.*—The dermis and epidermis appear normal. The only evidence of subcutaneous tissue is a small area in the H. and E. section consisting of sparse fat spaces embedded in connective tissue infiltrated with plasma cells, histiocytes, multinucleated cells and eosinophils—reminiscent of subcutaneous fat necrosis of the newborn.

*Blood picture.*—Hb 92% (13.7 grammes %); R.B.C. normal in appearance. Film not streaky. E.S.R. 3 mm. in the first hour. A number of atypical large mononuclears. Very suggestive of glandular fever. Total W.B.C. 14,000 per c.mm.:—neutrophils 5,000 (34%) no nuclear shift, eosinophils 1%, basophils 2%, lymphocytes 7,000 (50%), monocytes 3%, plasma cells 1%, atypical large mononuclears 1,300 (9%).

*Paul-Bunnell tests.*—18.3.53: Titre of unabsorbed serum: 1 in 56; titre of serum absorbed by ox cells: 1 in 56; titre of serum absorbed by guinea-pig kidney: 1 in 28. The titre, combined with the atypical absorption pattern, strongly supports the diagnosis of glandular fever.

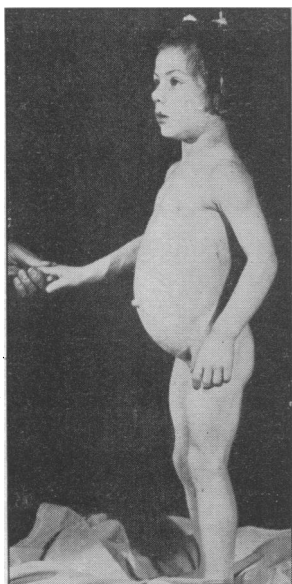


FIG. 1.

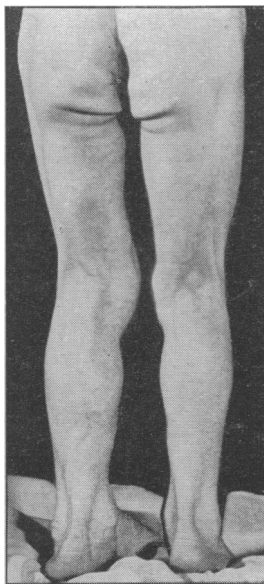


FIG. 2.

15.6.53: Titre of unabsorbed serum: 1 in 28; titre of serum absorbed by ox cells: 1 in 14; titre of serum absorbed by guinea-pig kidney: 1 in 14.

*Liver function tests.*—18.3.53: Blood urea: 35 mg.%. Thymol turbidity (MacLagen): 13 units (considerable increase). Serum colloidal gold (MacLagen) partial precipitation (4).

15.6.53: Serum thymol turbidity: 34 units. Serum colloidal gold: slight precipitation (2). Serum cholesterol: 250 mg.%.  
Cholesterol.—250 mg.% (slight increase).

*Urine.*—Twenty-four-hour intake 16; output 15 oz. Sp. gr. 1020. No sugar or other abnormal constituents.

*Blood sugar curve.*—Fasting level 68 mg.%; level at  $\frac{1}{2}$  hour 138; level at 1 hour 136; level at  $1\frac{1}{2}$  hours 76; level at 2 hours 68.

Dose of glucose 17.5 g. All specimens of blood were lipæmic.

X-rays: Chest—trachea deviated to the right. Bones and skull normal; bone age  $4\frac{1}{2}$  years.

There are only 4 previously reported cases of generalized lipodystrophy in children (Feer, 1915; Weber and Gunewardene, 1919; Hansen and McQuarrie, 1940; Corner, 1952). This case is of interest because of the association of abnormally rapid growth with disappearance of the subcutaneous fat depots. The term lipodystrophy is used advisedly: the fact that the child has no muscle wasting, and is a normal weight for her over-average height, the failure to demonstrate the presence of any subcutaneous fat in a satisfactory biopsy specimen, and the absence of facial subcutaneous fat, which persists in wasting diseases, making it very unlikely that the condition is due to inanition; but it is employed descriptively rather than as the name of a disease entity.

*Comment.*—It was suggested during the discussion that the patient might belong to the group described by Dr. Alexander Russell (1951) at the British Pædiatric Association meeting in April 1951 under the title of the diencephalic syndrome. Dr. Russell's cases had features (generalized wasting, advanced bone age, and temporary acceleration of growth) in common with the child described here, but also others (tendency to hypoglycæmia, peculiar temperament, low blood pressure, diabetes insipidus, signs of intracranial tumour) which she lacks. There is no evidence which makes it possible either to confirm or to deny the suggestion. Dr. Russell, who has now seen the child, thinks it likely that she has a diencephalic lesion, probably a glioma—and she will be followed with that possibility in view, but full air studies have failed to reveal such a lesion.

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