ARACHNODACTYLY IN A BANTU CHILD

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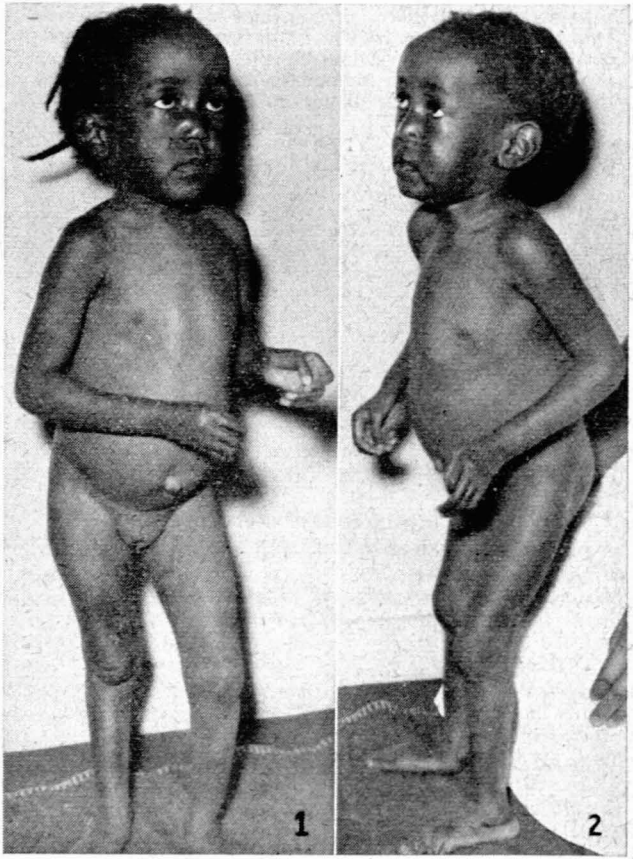
Marfan's syndrome is a rare anomaly, especially in the Bantu. A case of this condition in a Bantu child is submitted.

Samaria (Figs. 1 and 2) is a Bantu child, 2 years old. She presents anomalies met with in arachnodactyly (synonyms: Marfan's syndrome, dolichostenomelia, hyperchondroplasia). She was born on 5 May 1955, full time. Pregnancy and labour were normal. She was of average birth weight, breast fed 9 months, crawled at 6 months, and walked at 13 months. She has been delicate since birth.

Samaria was admitted to the Germiston Hospital on 10 May 1957 to determine her failure to gain weight, poor appetite, and perpetual tiredness. Her features are remarkable. She is tall, thin and lanky. Her height is 35 inches (average 32 inches), her span 37 inches (average for height 35 inches). Her weight is 22½ lb. (average 26 lb.). She has a sad, elderly appearance and looks twice her age. Her extremities are long and slender. The head is long and narrow (Fig. 4). There is bossing of the frontal bones. The facial bones and lower jaw are poorly developed. Her lips are thinner than in the average Bantu child. Her nose is large, broad and flattened. Her ears are prominent (Fig. 4), with deficient cartilage. Her palate is highly arched. She is mentally retarded.[1] She makes no attempt to speak. Her disposition is pleasant and peaceful, and she is happy in the company of children. Her walk is slow, awkward, unsteady, with a distinct forward bend. She falls easily when walking. She is always tired, and is usually found sitting in a sagging, slouched posture (Fig. 3). There is diminished subcutaneous fat. Her hand grip is feeble. Her hands and feet are remarkably long and narrow (Figs. 5 and 6). She has long, tapering fingers and toes, with hypermotility. There is ulnar deviation of the hands and valgus deformity of the feet. The thoracic cage shows marked asymmetry. She is pigeon-chested, with pronounced precordial bulge of the left side and flattening of the right. There is dorsal kyphoscoliosis. Blood count, blood chemistry, and blood pressure are within normal limits.

Eyes. There is a mild degree of epicanthic folds. The pupils are small and contracted and show limited reaction to atropine. The irides show a partial ectropion of the pigment border. Iridodonesis (tremulous irides) is present, and subluxation of the crystalline lenses.[2] Despite these defects her eyesight is fairly good.



Figs. 1 *and* 2. Author's case. Girl aged 2 years; looks older. Note stance, pigeon chest, tall, slender long extremities. Hands and feet long and narrow. Long, thin fingers and toes, and valgus deformity of feet. Note peculiar stare.

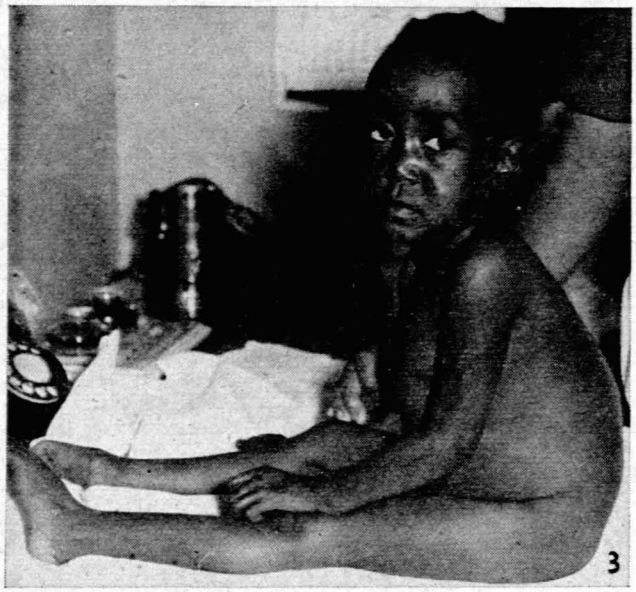


Fig. 3. Note tired look, slouched attitude, flat nose.

Cardiovascular System.[3] Examination revealed morbus cordis of the acyanotic variety. The heart sounds are abnormal but the congenital anomaly cannot be pin-pointed. The heart is displaced and a systolic pulmonary click is heard, suggesting some degree of pulmonary dilatation.

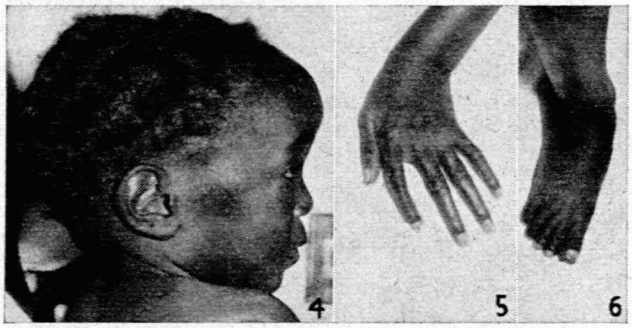


Fig. 4. Profile of head showing dolichocephaly.

Fig. 5. Note long, narrow hand with spider fingers and ulnar deviation.

Fig. 6. Note long, narrow foot with valgus deformity.

Radiological study showed a marked scoliosis of the umbar and dorsal spine. The mediastinum and heart were displaced towards the left side of the chest and the cardiac outline was distorted by rotation. The barium swallow was normal. The lung fields showed normal broncho-vascular markings.

DISCUSSION

Arachnodactyly is a hereditary condition transmitted by either sex. It is considered to be due to congenital mesodermal and ectodermal dystrophy suggestive of a genetically transmissable causal factor, possibly due to a defect in one or more chromosomes. It is doubtless determined early in embryonic life. It was first described by A. B. Marfan of Paris in 1896.[4] The basic features of this syndrome are fine, elongated, delicate extremities with tapering fingers and toes picturesquely named by Marfan as 'dolichostenomelia'. The name 'arachnodactyly' merely suggests a localized abnormality of hands and feet.[5]

The mosaic of this syndrome is inherited in varying degrees. Doubtlessly many atypical cases and *formes frustes* escape recognition—'incomplete' forms of arachnodactyly.

The thoracic cage of our little patient is distorted. The chest is pigeon-shaped and the trunk shows kypho-scoliosis, possibly due to excessive longitudinal growth of the ribs resulting in projection of the sternum and twisting of the spine.

It was impossible to investigate heredity and familial factors in the case under review.

There is a definite tendency toward a centrifugal increase in the length of all the long bones of the body. Hence the extraordinary length of the fingers and toes, so descriptively named 'spider fingers', 'spider toes'.

The Prognosis depends on the cardiac lesion. There is a high mortality in childhood with a marked tendency to respiratory disease, these patients being highly susceptible to intercurrent infection, especially of the lower respiratory tract. Samaria contracted measles during her stay in hospital in July 1957. She was *in extremis* and, but for the antibiotic cover, it is doubtful if she would have survived.

Treatment is symptomatic.

SUMMARY

A case is described of a 2-year-old Bantu female with symptoms of arachnodactyly. She is tall, thin and lanky, with 'spider hands and feet,' muscular dystrophy, deficient subcutaneous fat, laxity of ligaments and awkward gait. The thoracic cage is distorted and the trunk shows kypho-scoliosis. She is mentally retarded. Ocular anomalies are present. She has an acyanotic congenital heart.

The general features of this disease are briefly discussed.

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