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Bourneville's tuberous sclerosis: "When the cat is away . . ."

Whilst deputising for Mr L J F Delasiauve at the Salpêtrière on 18 July 1867, Désiré Magliore Bourneville (1840-1909) observed a child with the syndrome which now bears his name. He published the case promptly¹ and thereby his name gained eponymous precedence over Dr Hartdegen who made similar observations at the same time.² Bourneville's patient was L Marie aged three, born of parents without neurological abnormality or consanguinity.

" . . . breast fed by a wet nurse until 14 months. During this time she may have had several convulsions restricted to the eyes. At two years seizures appeared, during which the arms shook and turned slightly. It was particularly in the head. Marie never walked or talked. She gradually deteriorated...a hopeless case.

The eyes are dull . . . Rosaceous and pustular acne(sic) of the face; a confluent vesiculopapular rash over the nose, cheeks and forehead; numerous small mollusca on the nape of the neck, which is abnormally short. . . The right arm is paralysed, not complete. . . The left leg is longer and heavier than the right. The right thigh is adducted and flexed on the pelvis; the foot is flat, in varus, and violaceous. . . The legs are bent and crossed in bed. Constant dribbling. . . The seizures came in series. . . April 20th-30th fits. . . enema of bromide of camphor 2 grains."

After further fits the child died on 7 May at 3 am. At necropsy the brain weighed 1000 gm. He saw lesions: "rounded islets, forming protuberances of vari-

able size. They are whitish and opaque. . . a sort of hypertrophic sclerosis of various parts of the convolutions. The cavity of the lateral ventricle is normal. Islets. . . in frontal. . . temporal convolutions... corpus callosum, hippocampal convolution, Ammon's horn, paracentral lobule, corpus striatum. The right kidney showed three whitish mamillary hard masses projecting 3-5 mm. One is the size of a walnut. In addition 15 other small masses. . . when cut have the appearance of cancerous tissue. The left kidney shows similar but less numerous lesions.

The origin of this illness is obscure."

Bourneville clearly described the mental retardation, adenoma sebaceum, and the malformed sclerotic tubers of the brain. He subtitled the case "Tuberous sclerosis of cerebral convolutions: Idiocy and hemiplegic epilepsy." He described similar tubers in the kidneys, though he thought the latter appeared cancerous. He recorded both focal and clonic convulsive fits. The initial fits "especially the head" may resemble infantile spasms, but there were plainly focal and convulsive elements. And, it is likely that the child had an infantile pattern of right hemiparesis, though flexion and adduction of both arms and legs is described. He does not mention spasticity or reflexes.

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1 Bourneville, Contribution a l'étude de l'idiotie. *Archives de Neurologie* 1880;1:69-91.

2 Hartdegen A. Ein fall von multipler Verhärtung des Grosshirns nebst histologisch eigenartigen harten Geschwülsten der Seitenventrikel ("Glioma gangliocellulare") bei einem Neugeborenen. *Arch Psychiat* 1880;11:117-31.