

Although the progression of symptoms did not seem to shorten life, the crippling skeletal deformity, poor vision, and bilateral nerve deafness caused severe debility early in life with concomitant diminution of earning capacity and lowering of social status.

A mild diabetes insensitive to insulin was present in 3 cases. In one case, which has already come to autopsy, bilateral adrenal hyperplasia accompanied by basophilic hyperplasia of the pituitary was found. Endocrine study in the other patients studied, however, failed to disclose any endocrine abnormality despite the gross hyperplasia of the adreno-hypophyseal axis.

The above defects appeared to be inherited as a dominant trait. The ratio of the affected to non-affected individuals was approximately 1 to 1, i.e. about 50% of the family were affected. There was evident failure of the trait to skip a generation and complete absence of consanguinity.

The similarity and differences between this syndrome and the other syndromes previously mentioned will be discussed. In one patient, for example, the clinical picture at the age of 17 was that of Werner's syndrome, but now 35 years later he shows all the features of Refsum's syndrome. The overlapping clinical findings in our family with the three syndromes mentioned suggest a broad spectrum of neuroectodermal defects, which may include these and certain other related conditions as varying manifestations of a common underlying dysplasia of the neuroectodermal system.

221. GERSTENBRAND F., LORBECK W. and FOITL G.
(Vienna, Austria) Results of neuro-psychiatric
examinations in congenital heart disease

Thirty-two patients of the Second Surgical Clinic of the University of Vienna, all suffering from congenital heart disease, were subjected to a neurological, psychiatric, EEG and psychological examination.

The anomalies were the following:

Fallot's syndrome	6 cases
Atrial septal defect	6 cases
Pulmonary stenosis	7 cases
Patent ductus	5 cases
Aortic valve stenosis	4 cases
Mitral stenosis	2 cases
Eisenmenger's syndrome	1 case
Tricuspid atresia	1 case

Classification according to severity of the disease was established:

1. severe cases with cyanosis at rest and polycythaemia
2. severe disease with cyanosis during exertion
3. moderate
4. mild

The average age of the patients was 21 years. There were 16 males and 16 females. Manifest neurological signs were only found in 3 patients: a focus in the region of the medial cerebral artery in 2 cases, in one of them only transient, and athetoid-choreiform hyperkineses in the third. Two of these cases were in the group with cyanosis.

Four patients gave a history of epileptic attacks. One of these had temporal fits, but the routine EEG in this case was normal.

The percentage of neurological deficiencies in our series - including the cases with epileptic attacks - was therefore 30.

In 7 cases Adams-Stokes attacks were observed, in 8 cases hypoxia symptoms resembling high altitude disease were described. Three patients had bouts of headache.

The total number of patients who showed cerebral complications was 22, i.e. 67%. No correlation with the underlying cardiac disease could be detected, but the com-

plications were mainly found in the group with cyanosis and in the severe cases without cyanosis.

EEG changes were found in 16 of 30 examined patients and consisted of generalized changes, focal changes or paroxysms. Again the cases with cyanosis had the highest percentage of pathological EEGs.

Following operation under hypothermia with temporary circulatory occlusion in 7 cases there was no deterioration of the EEG.

In 26 cases a psychological examination was performed in the form of an efficiency and personality test. Almost all the patients showed neurotic changes and in 14 a certain degree of debility was found. Reduction signs in the sense of dementia were seen in 20 patients and in 3 further cases this was found on clinical examination. Again, a striking frequency incidence was found in the group of cyanotic patients (11 of 14). It is our opinion that these reduction signs are caused by the chronic hypoxia, as of 15 cases with cyanotic heart diseases 14 showed hypoxic damage. Six patients showed pseudo-dementia.

Fourteen of the total 32 cases had various other malformations; 7 of these showed an abortive Friedreich's syndrome.

In our material therefore almost all the cases of congenital heart disease showed some form of cerebral damage and almost half of these cases had other malformations.

222. GRANONE F. (Vercelli, Italie) Nature et mécanisme d'action de l'hypnotisme

Je pense que de nos jours encore la nature intime de l'hypnotisme n'est pas tout à fait éclaircie.

Une synthèse de nos connaissances actuelles en ce domaine, nous conduit à formuler quatre théories fondamentales, dans lesquelles se trouvent comprises, à mon avis, toutes celles décrites depuis le début du 18ème siècle.

1. La théorie sensualiste physique.
2. La théorie psychique subjectiviste.
3. La théorie psychanalytique.
4. La théorie réflexologique.

En ce qui concerne la première théorie il faut remarquer que l'existence d'une force humaine particulière, capable d'induire une transe, doit être encore démontrée même si l'on est porté à l'admettre, du fait de la facilité différente avec laquelle divers hypnotiseurs peuvent induire une transe immédiate chez le même sujet, du fait aussi qu'on peut reproduire certains états neuropsychiques de la transe par des moyens purement physiques et donc sans suggestion mentale, et que ces états (première phase de l'hypnose) sont identiques chez l'homme et chez les animaux soumis à certaines stimulations physiques et sensorielles.

A l'appui de la deuxième théorie on peut remarquer qu'une grande partie de la phénoménologie hypnotique peut apparaître par suggestion ou par autosuggestion, mais il n'en reste pas moins qu'il existe certains symptômes psychiques et somatiques neurovégétatifs qui sont tout à fait différents et peuvent être considérés comme caractéristiques de l'hypnose.

Enfin l'hystérie et l'hypnotisme ne peuvent guère être mis sur le même plan, plusieurs considérations permettant d'écarter cette hypothèse.

Les interprétations purement physiologiques, selon la doctrine des réflexes conditionnels ou les doctrines psychanalytiques, peuvent apporter une contribution importante à la connaissance de l'hypnotisme mais ne sont pas en mesure d'éclaircir la genèse, la nature et le dynamisme de tous les phénomènes hypnotiques.

Le mécanisme d'action de l'hypnose se fonde sur l'interdépendance des phénomènes somatiques, psychiques et viscéraux et sur des concepts fondamentaux d'unité psychosomatique.

A la lumière des connaissances actuelles on doit admettre que les niveaux du névraque, sièges de la corrélation psychosomatique en hypnose, sont le cortex céré-

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