M. Minauf: 
Die sogenannten amaurotischen Idiotien
Veröffentlichungen aus der Pathologie, Heft 96
Fischer, Stuttgart 1975
VIII + 112 pp., 25 fig., 1 cpl., 3 tab.; DM 48.-
ISBN 3-437-10380-6

This monograph by Margaret Minauf is devoted to amaurotic idiocies. It aims at reclassifying these diseases in the light of recent ultrastructural and biochemical findings. Especially the latter, beginning with Klenk’s analysis in 1939, made the old subdivisions based on age and eponymic designations unnecessary and irrelevant. Pathogenetical classification, accordingly, is still required.

One example of nosological difficulty is ‘infantile amaurotic idiocy’. Until recently it was considered to be a single disease, but is now known to be a disease group. It consists of several kinds of GM2 gangliosidoses with different hexosaminidase deficiencies, of GM1 gangliosidoses caused by a defect in /β-galactosidase as well as of cases of ceroid lipofuscinosis. Juvenile and adult cases form an inhomogeneous morphological group according to the same criteria. Further Section

In the first part of her work, the author describes her own case material on ‘amaurotic idiocy’, of which two are infantile, two juvenile, two late infantile and one adult. Each case was thoroughly studied clinically (including ophthalmologically), morphologically (including ultrastructure) and biochemically. The morphological findings of her own cases as well as those of others are richly illustrated and the author emphasizes the variability of the morphological findings in each age group, e.g. some of the late infantile cases are cases of GM1 or GM2 gangliosidoses and still others are ceroid lipofuscinosis with typical curvilinear bodies.

In the second part of her monograph, the author discusses an astonishing amount of neuropsychological, ophthalmological, morphological and biochemical data for each age group. She also reviews the rare congenital cases described in the literature and all those which still remain difficult to classify.

In the final part of her publication, she reclassifies the cases according to biochemical criteria. Two large groups can be formed: gangliosidoses and ceroid lipofuscinoses. The former can be subdivided into those with a defect in hexosaminidases (GM2 gangliosidoses) and those in which the galactosidases are lacking (GM1 gangliosidoses). The GM1 gangliosidoses can be further subdivided into variants B, A and AB according to the still present, nonreduced hexosaminidase. The author then discusses the degree to which the former eponymic designations and age groups correspond to these new classifications.

The feasibility of genetic counselling is discussed and the necessity of a multidisciplinary approach to this disease group is stressed. The extensive documentation of the publication and its large-scale approach to clinical pathology make it a work of reference, especially for pediatrists, neuropathologists and neuro-chemists.

A. Probst

E. Riigheimer und D. Heitmann: 
Die Neuroleptanalgesia — Bilanz einer Methode
NLA-Workshop Bad Rappenhall, 1974
Thieme, Stuttgart 1975
X + 195 pp., 58 fig., 33 tab.; DM 39.-
ISBN 3-13-5346-01-3

Nearly 15 years after the inauguration of neuroleptanalgesia (NLA), a large number of competent experts from the Federal Republic of Germany, Austria and Switzerland met to compare the performance of NLA with that of conventional anaesthetics.

Discussions about fundamental problems as well as problems of application were based on conceiving ‘pain’ as a result of three analytic steps: (1) localizing, (2) identifying, and (3) experience. Drugs derived from the butyrophenons (neuroleptic component of NLA-effect) and other derivatives from the synthetic morphine-like pain killers (analgesic component of NLA-effect) are presented.

Representing operative specialities, the experts critically weigh reports concerning the NLA standard technique and its variations - not omitting particularities of NLA in children and in the aged. Out of its abundance, it is made evident that NLA has its fixed position in the methodology of operative diagnostic and therapeutic procedure, and that it offers a lot of elbowroom for the anesthesists to be able to meet the patients’ needs as well as the surgeons’ claims. To give an example in neurology/neurosurgery, NLA offers the advantage of lowering raised CSF pressure, stabilizing circulation, providing ideal tolerance of tube, and rapid recovery postoperation; optimal conditions for supervision as no other comparable procedure has been able to offer before. Sporadical side effects, namely, necessity of intubation and ventilation of the patient, shivering (like in halothane), the ‘living’ of the operation as well as a questionable hypersensitivity to procainamide scarcely limit the practical usefulness of this method.

Actual contributions about postoperative analgesia, and NLA-drugs in intensive care, round up the wide range of excellently-documented extensive reports. The editors E. Riigheimer and D. Heitmann present a balance which answers not only retrospective but also prospective questions.

A.R. Koch, Basel

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