described by Tans[11] in a case of OPCA, concluding that the brain-stem lesions were probably not secondary to the cerebellar anomaly. It is obvious that Norman and Urich's case had not a pure ponto-neocerebellar hypoplasia but a neocerebellar hypoplasia with multisystem degeneration that could be included under the general category of OPCA. Similar cases have recently been published.[12-15] Pathogenetically, most authors favour the occurrence of a developmental anomaly producing cerebellar hypoplasia and predisposition of some neuronal systems to abiotrophy.[2 3 12-14]

Finally, I agree with the statement that OPCA is a common pathological end point for several clinically and genetically distinct disorders.[1] In fact, olivopontocerebellar lesions have been described in a patient with adrenoleukodystrophy.[16]

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Harding and Erdohazi reply

Dr Berciano takes issue with us[1] for disagreeing with his classification[2] of two particular case reports, Case 1 of Norman and Urich[3] and that of Gross and Kaltenbäck[4] as olivopontocerebellar atrophy (OPCA). His letter highlights the particular problems of morphological classification with childhood cerebellar syndromes. There is certainly no unanimity in the literature. Urich[5] classified both cases with pontoneocerebellar hypoplasia (PNCH). Koeppen and Barron[6] also describe Norman and Urich's case as cerebellar hypoplasia, while Friede[7] in his encyclopaedic treatise expresses some difficulty over the interpretation of this case.

The case of Gross and Kaltenbäck would be rather unusual for OPCA in view of the histologically normal cerebellar cortex and severe neuronal loss from the dentate and red nuclei and degeneration of the superior cerebellar peduncles. In a recent review Koeppen and Barron[6] remark that Purkinje cell loss is the outstanding feature in the cerebellum in OPCA, and can be so sweeping that none are found, while deep nuclei and their efferents are generally well preserved. Although its extent can be quite variable cerebellar cortical involvement is found in almost all cases.[6 8] On the other hand dentate involvement is much less frequent, about a third of cases in Berciano's review,[2] and this is usually only gliosis; the dentate cells and superior cerebellar peduncles are generally well preserved.[8] This is not surprising if one views OPCA, as Berciano suggests, as a linked degeneration commencing in the pontine nuclei with later transsynaptic degeneration of the cerebellar cortex. Dentate degeneration would presumably follow from loss of Purkinje axon activity. Indeed the few well defined reports of OPCA with "normal cerebellar cortex" also have normal dentate nuclei.[9-13]

Berciano is correct in thinking that Norman and Urich's case is a form of cerebellar hypoplasia, although it is not so obvious as he avers (see Friede[7]). But to lump cases of PNCH with OPCA is to confuse fundamentally different processes. In OPCA, pontine and arcuate nuclei and the inferior olives are the prime targets for degeneration. In the rare condition PNCH there is severe *hypoplasia* of the neocerebellar cortex and pontine nuclei and usually a remarkable malformation of the dentate nuclei which are broken into a series of islands. These classical findings[14 15] are confirmed in recent papers[16-18] and in several personally examined cases. The unexpected finding in our familial cases[1] of cerebellar pathology completely different from PNCH, but quite typical of OPCA and yet occurring in the neonatal period is what prompted our publication. We have examined a third unrelated case with a very similar clinical and pathological picture, which we are preparing for publication. In addition, it has recently come to our attention that two siblings with a very similar clinicopathological picture have been reported by Agamanolis *et al.*[19]

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Acute dystonic reaction with asterixis and myoclonus following metoclopramide therapy

Sir: We read with interest the letter of Chin-Song Lu and Wai-Shin Chu[1] about a patient who developed myoclonus and asterixis following treatment with metoclopramide. The authors did not find any cases described of these complications of metoclopramide therapy.

In 1984 our group reported a 69 year old woman who developed generalised and irregular asymmetric muscle jerking movements mainly of the right side, 24 hours after the first oral dose of 10 mg of metoclopramide (total dosage: 40 mg).[2] Examination revealed a right hyperreflexia and extrapyramidal rigidity. CT showed cerebral, cerebellar and brainstem atrophy. The myoclonus cleared up 24 hours after discontinuation of metoclopramide. During follow up examinations, 6 months and 1 year later, the patient was found to be normal.

In our patient we believe that myoclonus could be attributed to the metoclopramide action on a hypothetic subclinical disturbance of dentatorubral pathways due to cerebellar and brainstem atrophy.

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Accepted 12 September 1988.