

# Right Cerebellar Hemisphere Agenesis in a 19-Year-Old Patient Presenting Behavioural and Thought Disorders: A Case Report

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## Abstract

*The origins of schizophrenia remain unclear, in spite of approaches based on neurobiological models. Links between these models and semiological aspects are sometimes not obvious because of clinical concomitances. We report here a case of right cerebellar hemisphere agenesis in a 19-year-old male patient presenting behavioural and thought disorders. This patient, who was adopted at the age of 2 ½ years, also presented very poor WAIS-R standard scores. This clinical case shows similarity with Andreasen's concept of "cognitive dysmetria" and, considering the major cognitive impairment of that patient, the possibility of what Kraepelin in 1919 termed "Pfpopschizophrenie" might be discussed. (German J Psychiatry 2002;5:70-74).*

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## Introduction

The psychopathology of schizophrenia has yet to be fully explained, in spite of approaches based on neurobiological models. Psychiatrists are confronted by semiological variability, and the link between models and clinical aspects is not obvious. We report on a case of a right cerebellar hemisphere agenesis in a 19-year-old male patient who was admitted for behavioural and thought disorders.

## Case Report

Mr X, 19 years old, was admitted at the request of his parents after a fugue that lasted 2 weeks, ending about 500 miles away from his home. Complaints included the fact that he appeared repetitive and his apparently uncontrollable attempts at travelling without a known destination, a tendency towards solitude, confabulations, and out-of-context initiatives.

## At Admission

The patient was quiet, had a good general appearance, and was cooperative. His speech was superficial, made up of sentences with fantastic contents and sometimes incoherent. He described the course of and his activities during his flight: He hitchhiked and although he had neither money nor driving licence, he went to see a car salesman with the aim of acquiring a car. Mr X did not present ideas of reference but complained of hearing the voice of one of his family members who had died many years ago. He also complained of losing control of his ideas: "All of a sudden it overflows, my head is overflowing, and I do not have control of anything anymore". There was no evidence of substance abuse, his mood seemed neutral, there was no anxiety and he oriented himself well in time and space. His parents were very anxious.

## Biography

Mr X was adopted at the age of 2½ years by a couple who were both 44 years old. In this family, he joined a 6-year-old boy who had already been adopted before him. Through his

paediatric file we found that he had been followed up from the age of 6 to 10, due to school retardation, notably in reading. He also presented with ataxia and apraxia. A CT-scan done at the age of 8 revealed that Mr X had a cerebellar anomaly. After 10 years' training on mechanics in a medico-educational institute, he left without qualification. During this period, his relatives noticed his tendency towards solitude and retreat. There followed a series of unusual incidents, such as the destruction of his own documents and of toys belonging to other children. His attitude became less and less understandable, culminating in his unpredictable and unmotivated running away from home.

## Physical Examination

Mr X did not present any disturbance of stature or walking. The neurological assessment for "soft" signs using the Neurological Evaluation Scale (Buchanan & Heinrich, 1989) revealed minor abnormalities in the following subtests: Tandem Walk (two missteps after completion of first full step); Audio-Visual Integration (one error); Stereognosis (one error); Fist - Ring test (difficulty in developing and maintaining a smooth, steady flow of movement); Rhythm Tapping Test (error in number of taps); Finger - Thumb Opposition (no major disruption of motion); Right - Left confusion (with one error). He presented major abnormalities in: Romberg Test (he stepped to maintain balance); Adventitious Overflow (he had irregular fluttering movement extended to hands); Tremor (marked); Graphesthesia (more than one error); Ozeretski Test (major disruption of movements); Memory (he remembered just three words); Rapid Alternating Movements (more than three mistakes in hand placement); Synkinesis (movements of his head even when told to keep head still). The following subtests did not reveal any abnormality: Cerebral Dominance (hand and foot dominance on the right); Convergence; Gaze impersistence; Finger to Nose Test; Glabellar Reflex; Snout Reflex; Grasp Reflex; Suck Reflex by placing a tongue depressor between his lips.

Laboratory test did not show any abnormalities.

To begin with, we suspected that Mr X was suffering from an insidious form of a chronic psychotic process. We prescribed an atypical neuroleptic drug as well as activities within the ward.

## Course

During the first weeks of his stay, Mr X collected miscellaneous materials (pebbles, papers, tumblers used for distributing tablets, etc.) and kept them in his room. He caused damage to the premises and showed little compliance with disciplinary rules. He seemed disconnected from reality. Further, he expressed morphological preoccupa-

tions: Mr X complained sometimes about his ears, describing them as deformed, soft.

After 3 months' follow-up, we noticed a decline in his desire to run away, but associated with decreased motivation, ambivalence, and poor interest in activities. His mood remained neutral most of the time.

In view of his functional disorganisation, we decided to perform additional examinations. The WAIS-R test (Wechsler's Intelligence for Adults - Revised) was given to Mr X. In the Verbal Scale comprising Information, Similarities, Arithmetic, Vocabulary, Comprehension, and Digit Span, he obtained scores which allowed us to establish a standard score of 25 for this subscale. In the Performance Scale, which comprises Picture Completion, Picture Arrangement, Block Design, Object Assembly, and Digit Symbol, he obtained a standard score of 23. Taken together, this gave him a test standard score of 48, which corresponds to a severe mental impairment. In spite of his repeated failures, he declared after this test that he had done well.

A nuclear magnetic resonance examination (Figures 1-4) conducted in various different planes without and after injection of gadolinium showed: At the posterior fossa level, an image occupying the right cerebellar lobe compatible with a liquid collection with a complete atrophy of the right cerebellar hemisphere. No abnormality was revealed at the level of the left cerebellar lobe and the vermis. A discrete asymmetry of the cerebral hemispheres was found in favour of the right. We did not find any expansive process, nor an abnormality of median structures or pathological deposits of gadolinium. The ventricular system was not dilated and there were no pathological findings in the periventricular white matter.

## Discussion

We diagnosed Mr X as having an insidious onset of a psychotic process such as schizophrenia, in accordance with DSM IV criteria (disorganisation of speech, behavioural disorders, apragmatism, social and active dysfunctions). With these elements, one would also consider the diagnosis of schizotypal personality disorder. This case may well suggest many other possibilities; at times, Mr X showed a lack of moderation with megalomaniac tendencies in his speech and actions, and a flight of ideas contrasting with apragmatism. In spite of his neutral mood, we did not exclude a current bipolar disorder. The history of Mr X revealed retarded development in childhood, with difficulties at school, confirmed by his poor WAIS-R standard scores. These elements may be related to his cerebellar malformation, but are also consistent with what Kraepelin in 1919 termed "Pfpopschizophrenie" (Doody et al., 1998). He estimated that around 7 % of all cases of dementia praecox arose in individuals with premorbid cognitive impairment.

Figure 1. Magnetic Resonance Tomography, Frontal Scan.



Figure 2. Magnetic Resonance Tomography, Left Cerebellum

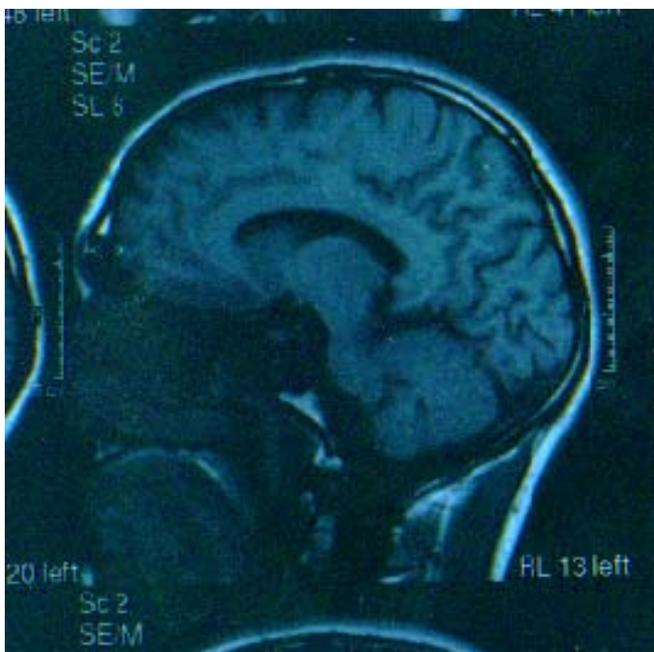


Figure 3. Magnetic Resonance Tomography, Right Cerebellar Lobe

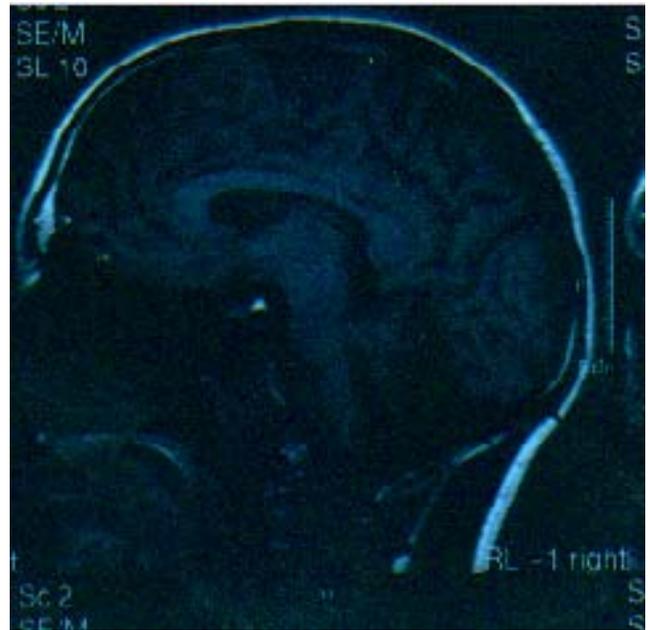
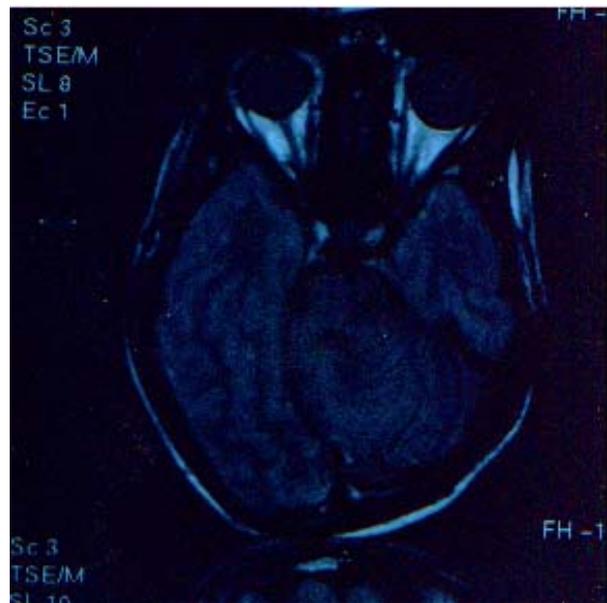


Figure 4. Magnetic Resonance Tomography, Caudal Scan



Another interesting aspect of this case lies in the concomitance of an abnormality of the cerebellum and psychiatric symptoms. Many authors have stated that the cerebellum probably plays a role in psychotic processes and several papers have described morphological and neurobiochemical abnormalities of this organ in schizophrenics.

Andreasen's (1996) hypothesis termed "cognitive dysmetria" stated that it is a neurological abnormality, implying that the cerebellum would be responsible for a disturbance of mental coordination at the origin of schizophrenia. It was suggested that this takes place by means of func-

tional changes in the neuronal circuits connecting the cerebellum to prefrontal and thalamic structures. From Microarray technologies (Mirnics et al., 2001) applied to the analysis of human brain disorders, a hypothesis has recently been formulated to suggest that schizophrenia is a disease of the synapse.

Lu & Yeh (2001) reported very recently the case of a patient in whom the ablation of a posterior fossa tumour involving the cerebellum was followed by the occurrence of schizophrenic symptoms.

Eastwood et al. (2001) measured the expression of three proteins (synaptophysin, complexin I, complexin II) at the synaptic level in the cerebellar cortex of sixteen schizophrenics, compared with sixteen normal subjects. In the schizophrenics, they found a decline of the mRNA of synaptophysin, complexin II and its mRNA, which brought them to conclude that schizophrenics have a cerebellar synaptic abnormality which is liable to alter the functioning of the neuronal circuits.

Melchitzky et al. (1999) reported evidence in macaque monkeys of the presence in certain lobules of the cerebellar vermis of dopamine receptors essentially at the level of granular layers, suggesting the potential role of this substance in certain cerebellar functions in primates.

Volz et al. (1999) conducted measurements on the nervous system of a group of schizophrenics compared with normal subjects by nuclear magnetic resonance and found a significant reduction in volume of the left cerebellar hemisphere and the right vermis in the schizophrenics, a reduction likely to be increased by alcoholism (Katsetos et al, 1997).

In the context of the neurodevelopmental model, it is suggested that impaired mechanics of synaptic transmission in specific neural circuits during childhood and adolescence ultimately result in altered synapse formation or pruning, or both, which manifest in the clinical onset of the disease. Most of the authors (Tibbo et al., 2000; Slater et al., 1998) found abnormalities associated with the frontal, temporal lobe and the cerebellum.

## Conclusion

A convergence of observations from clinical, neuroimaging and anatomical studies has focused attention until recently on the prefrontal cortex as a major area of alterations in schizophrenia. The concomitance in this young patient of a cerebellar abnormality, severe learning disability, and schizophrenic symptoms suggests that previous hypotheses need to be re-evaluated. Our understanding of the psychopathology of schizophrenia depends on it. The clinical assessment of people with schizophrenic symptoms should pay particular attention to screening for neurologi-

cal "soft" signs. Refined neuropsychological testing should also be done and imaging of the cerebellum performed.

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