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CONGENITAL MALFORMATIONS

MUSIC AFFINITY IN WOLF-HIRSCHHORN SYNDROME

Investigators at Nihon University School of Medicine, Tokyo, Japan, report two patients with Wolf-Hirschhorn syndrome (WHS) who have a strong affinity for music, not previously described as a feature of this syndrome. WHS is a congenital malformation syndrome that results from deletion of the short arm of chromosome 4, and is characterized by a “Greek warrior helmet” appearance, growth retardation, developmental delay, muscular hypotonia, epilepsy, and a language disorder involving verbal communication. *Patient 1*, a 20-year-old woman with developmental delay, presented with status epilepticus with fever at age 9 months and followed by simple and/or complex partial seizures up to 8 times daily, triggered by fever or bathing. Brain MRI at 7 years of age was normal. Seizures were partially controlled with AEDs by 4 years of age and stopped at age 13 years. Her mother first reported an affinity for music at age 2 years 3 months. *Patient 2*, a 9 year-old girl was diagnosed with WHS soon after birth because of her craniofacial features, and was confirmed by chromosomal analysis. She sat at 4 years and stood with support at 9 years of age. She presented with status epilepticus with fever at age 5 months, followed by afebrile seizures and recurrent status with fever, finally controlled at 34 months. Brain MRI at age 33 months showed reduced white matter volume. Major dysfunctions included atrial septal defect, vesicoureteral reflux, clubfoot, and eating difficulty. She cannot understand or speak words and has autistic behaviors. Listening to music had a calming effect since infancy, and music therapy started at age 5 years was followed by improvement in communication skills (eye contact, attention, and vocalizations). (Arakawa C, Fujita Y, Fuchigami T, et al. Affinity for music in Wolf-Hirschhorn syndrome: Two case reports. **Pediatr Neurol** 2014 Oct;51(4):550-2).

COMMENTARY. Music therapy is used in the treatment of autism and in other behavioral and developmental disorders [1]. Williams syndrome (WS), resulting from deletion of the long arm of chromosome 7, has a much higher incidence than WHS, and an increase in musical interest and ability associated with WS is widely known [2]. A functional MRI study of the musical affinity of patients with WS shows an association with increased activation of the right amygdala [2].

Distinctive spectrum of epilepsy and electroencephalogram patterns in WHS.

Analysis of 87 patients (54 females, 33 males; median age 3.6 years; range 1-25 years) with confirmed 4p16.3 deletion and WHS syndrome found 93% developed epilepsy in the first 3 years of life. Seizure patterns were generalized tonic-clonic in 74%, tonic spasms in 18%, complex partial in 12%, and clonic in 7%. Seizures were triggered by fever in 73% and occurred in clusters and as febrile status epilepticus in 50% during the first 3 years of life. Atypical absence seizures accompanied by myoclonus of the

eyelids and hands occurred between 1 and 6 years in 33%. Distinctive EEG abnormalities were seen in 90%. Epilepsy was well controlled in 81%, improved with age, and stopped at a median age of 4 years 6 months [3].

The stereotypic electroclinical pattern of WHS is described as intermittent bursts of 2-3 Hz high voltage slow waves with superimposed spike and spike wave activity in centroparietal or parietotemporal areas, often elicited by eye closure, and associated with myoclonic jerks [4]. The electroclinical pattern of WHS characterized by discharges mixed with slow components on eye closure is similar to that described in Angelman syndrome [5]. Awareness of the clinical neurological syndrome and characteristic electroclinical findings in WHS and AS might help in early diagnosis and treatment of associated epilepsy [6].

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HEADACHE DISORDERS

NEUROSURGEON'S PERSPECTIVE ON NEUROIMAGING FOR HEADACHES

Neurosurgeons at Washington University School of Medicine, St Louis, MO, address the proposed utility and negative impact of guidelines proposed by the American Headache Society (AHS), in conjunction with the Choosing Wisely initiative of the American Board of Internal Medicine. The first of five recommendations approved by the AHS Choosing Wisely [1] Task Force, "Don't perform neuroimaging studies in patients with stable headaches that meet criteria for migraine," is the subject of this neurosurgical perspective. The Choosing Wisely initiative focuses on reducing health care spending by setting guidelines to limit tests ordered by physicians, and if adopted, the guidelines threaten to negatively impact the care and outcomes of patients with brain tumors.

Patients with brain tumors may present with isolated headaches in the absence of other neurological symptoms and signs. Despite classic teaching, headaches caused by tumors may be similar to tension-type or migraine-type headaches in 77% and 9% of cases, respectively. A review of 95 patients (10 children) diagnosed with a brain neoplasm by open biopsy at Washington University found isolated headaches were the only complaint in 11.6% of brain tumor patients, and 24.2% presented with isolated headaches, no symptoms, or only non-specific symptoms. If the Choosing Wisely Task Force [1] recommendations had been followed, diagnosis would have been delayed or missed for 3 of 11 isolated headache patients (3.2% of all brain tumor patients). If the American College of Radiology/Consumer Reports recommendations had been followed, "Don't do imaging for uncomplicated headaches," a diagnostic delay/error would have occurred for 7 of 11 isolated headache patients (7.4% of all patients). Under the proposed