Singing was associated with complex behavioral changes including euphoria, laughing, gestural automatisms, and dancing. Humming occurred in seizures affecting the temporal lobe, whereas singing was associated with seizures originating in the frontal lobe, particularly the right prefrontal cortex. (Bartolomei F, McGonigal A, Guye M, Guedj E, Chauvel P. Clinical and anatomic characteristics of humming and singing in partial seizures. **Neurology** July 31, 2007;69:490-492). (Reprints: Dr F Bartolomei, Service de Neurophysiologie Clinique, CHU Timone, 264 Rue de Pierre, 13005-Marseille, France).

COMMENT. Musicogenic epilepsy is well known (Critchley M. **Brain** 1937;60:13-27). Seizures occur on hearing or playing music, and often, in response to specific compositions. Laughter as a form of epilepsy (gelastic seizures) is also reported, usually in young children with hypothalamic hamartoma (Mullatti N et al. **Epilepsia** 2003;44:1310-1319) **(Ped Neur Briefs** Nov 2003;17:81-83). Singing as a musical automatism during seizures is uncommon, the above authors citing reports by Vidailher M et al. 1989, and Doherty MJ et al. 2002. The anatomy of music perception has been studied using a high-resolution PET scanner at the University of Caen, France, and the Wellcome Department of Cognitive Neurology, London, UK (Platel H et al. **Brain** 1997;120:229-243). The left hemisphere is dominant for rhythm, tune recognition, and pitch perception, whereas the right hemisphere subserves timbre or quality of tone perception.

## ROLE OF *ARX* GENE IN INFANTILE SPASMS AND DYSTONIA

The role of *ARX* gene in a syndrome of infantile spasms with generalized dystonia was investigated in 6 boys from 4 families at the University of Florence, Italy, and other centers in Italy, Japan, and USA. Mutation analysis in 115 boys with cryptogenic infantile spasms found an expansion of the first PolyA tract of *ARX* in 6 (5.2%) ages 2 to 14, from four families, including two pairs of brothers. All six had mental retardation and generalized dystonia that developed at 6 months and progressed to quadriplegic dyskinesia within 2 years. Three had recurrent status dystonicus. Multifocal small cavities in the putamina were seen on the MRI of 4 children. *ARX* gene testing is recommended in boys with infantile spasms and unexplained dyskinetic quadriplegia. (Guerrini R, Moro F, Kato M et al. Expansion of the first PolyA tract of *ARX* causes infantile spasms and status dystonicus. **Neurology** July 31, 2007;69:427-433). (Reprints: Dr R Guerrini, Clinical Pediatric Neurology, Department of Pediatric Neurosciences, University of Florence, and Pediatric Hospital A Meyer, via Bonvicini 62, 50132 Firenza- Italy).

COMMENT. Several brain malformations are associated with mutations of the *ARX* gene, including X-linked lissencephaly with abnormal genitalia, hydrocephalus, and agenesis of the corpus callosum with abnormal genitalia (Proud syndrome). Syndromes associated with the same gene and without brain malformations include X-linked infantile spasms, Partington syndrome (mental retardation with mild dystonia), and X-linked mental retardation. The authors describe an "infantile epileptic-dyskinetic encephalopathy" in males with expansion of the first PolyA tract of *ARX*, and consisting of severe mental retardation, early-onset infantile spasms, and severe progressive generalized dystonia with status dystonicus, some patients showing basal ganglia cysts.