

dopaminérgicos, bloqueadores cárnicos, anticonvulsivantes, manganeso, monóxido de carbono y metanol. Generalmente producen distonía focal aguda de rápida recuperación al suspender el fármaco. Sin embargo puede existir también distonía tardía.

Causas infecciosas/post infecciosas. La encefalitis por anticuerpos anti receptores NMDA, la panencefalitis esclerosante subaguda y la encefalomielitis pueden mostrar clínica de distonía aguda o subaguda.³

Psicógena. Cuando el movimiento anormal no es clínicamente clasificable, pero se asemeja a la distonía,(pseudo distonía) debe sospecharse causa psicógena, la que se confirma por negatividad de neuroimágenes y análisis toxicológico.

CONCLUSIONES

Para abordar en forma certera las distonías en pediatría debe realizarse primero descarte de causas secundarias, lo cual se realiza esencialmente con la anamnesis detallada, semiología clínica, estudios toxicológicos y de neuroimagen. Si la clínica más las neuroimágenes son concordantes con determinadas enfermedades metabólicas/neurodegenerativas deberá realizarse estudios metabólicos complementarios. En casos de distonía con normalidad de todos los exámenes antes mencionados debe plantearse la causa primaria y eventualmente realizar estudio genético. De esta forma se puede definir mejor el pronóstico y tratamiento de las distonías.

BIBLIOGRAFÍA

1. Oppenheim H. About a rare spasm disease of childhood and young age (Dysbasia lordótica progresiva, dystonia musculorum deformans). *Neurologische Centralblatt* 1911;1090-107.
2. Mink JW. Special concerns, defining, studying and treating dystonia in children. *Mov Disord* 2013;28(7):921-25.
3. Sanger TD, Delgado MR, Gaeble-Spira D, Hallett M, Mink JW. Classification and definition of disorders causing hypertonia in childhood. *Pediatrics*. [Revista en Internet].2003 [Consultado el 23 de febrero del 2014];111(1):e89-e97. Disponible en: <http://www.ncbi.nlm.nih.gov/pubmed/12509602>.
4. Albanese A, Bhatia K, Bressman S, De Long MR, Fahn S, Fung VSC, et al. Pheno-menology and classification of dystonia: a consensus update. *Mov Disord* 2013; 28(7):863-73.
5. Fernandez-Alvarez E, Nardocci N. Update on paediatric dystonia. *De-generative Neurologic Neuromuscular Disease* 2012;2: 29-41.
6. Uc EY , Rodnitzky RL. Dystonia in children. *Semin Ped Neurol* 2003;10(1):52-61.
7. Deonna TW, Ziegler AL, Nielsen J. Transient idiopathic dystonia in infancy. *Neuropediatrics* 1991;22(4):220-24.
8. Deonna TW, Ziegler AL, Nielsen J. Transient idiopathic dystonia in infancy. *Dev Med Child Neurol* 2006;48(2):96-102.
9. Deonna T, Martin D. Benign paroxysmal torticollis in infancy. *Arch Dis Child* 1981; 56(12): 956-59.
10. Giffin NJ, Benton S, Goadsby Bj. Benign paroxysmal torticollis of infancy: four new cases and linkage to CACNA1A mutation. *Dev Med Child Neurol* 2002;44:490-93.
11. Werlin SL, D'Souza BJ, Hogan WJ, Dodds WJ, Arndorfer RC. Sandifer syndrome: an unappreciated clinical entity. *Dev Med Child Neurol* 1980;22:374-8.
12. Ozelius LJ, Hewett JW, Page CE, Bressman SB, Kramer PL, Shalish C, et al: The early-onset torsion dystonia gene (DYT1) encodes an ATP-binding protein. *Nat Genet* 1997;17(1):40-8.
13. Blanchard A, Ea V, Roubertie A, Martin, M, Coquart C, Claustres M, et al. DYT6 dystonia: review of the literature and creation of the UMD Locus-Specific Database (LSDB) for mutations in the THAP1 gene.
14. Bentivoglio AR, Ialongo T, Contarino MF, Valente EM, Albanese A. Phenotypic characterization of DYT13 primary torsion dystonia. *Mov Disord* 2004;19(2):200-6.
15. Ichinose H, Ohye T, Takahashi E, Seki N, Hori T, Segawa M, et al: Hereditary progressive dystonia with marked diurnal fluctuation caused by mutations in the GTP cyclohydrolase I gene. *Nature Genetics* 1994;8:236-42.
16. Segawa M, Nomura Y, Nishiyama N. Autosomal dominant guanosine triphosphate cyclohydrolase I deficiency (Segawa disease). *Ann Neurol* 2003;33(3):195-201.
17. Nygaard TG, Waran SP, Levine RA, Naini AB, Chutorian AM. Dopa-responsive dystonia simulating cerebral palsy. *Pediatr Neurol* 1994;11:236-40.
18. Di Capua M, Bertini E. Remission in dihydroxyphenylalanine-responsive dystonia. *Mov Disord* 1995; 10(2):223.
19. Kurian MA, Li Y, Zhen J, Meyer E, Hai N, Christen HJ, et al. Clinical and molecular characterisation of hereditary dopamine transporter deficiency syndrome: An observational cohort and experimental study. *Lancet Neurol* 2011;10(1):54-62.
20. Asmus F, Zimprich A, Tezenas Du Montcel S, Kabus C, Deuschl G, Kupsch A, et al. Myoclonus-dystonia syndrome: Epsilon-sarcoglycan mutations and phenotype. *Ann Neurol* 2002;52(4):489-92.
21. Campistol J, Cusi V, Vernet A, Fernández-Alvarez E. Dystonia as a presenting sign of subacute encephalomyopathy in infancy. *Eur J Ped* 1986;144(6):589-91.
22. Morava E, Steuerwald U, Carrozzo R, Kluijtmans LA, Joensen F, Santer R, et al. Dystonia and deafness in children with SUCLA 2 defect; clinical course and biochemical markers in 16 children. *Mitochondrion* 2009;9(6):438-42.
23. Burke RE, Fahn S, Gold AP. Delayed -onset dystonia in patients with "static" encephalopathy. *J Neurol Neurosurg Psychiatry* 1980;43(9):789-97.
24. Cerovac N, Petrovic I, Klein C, Kostic VS. Delayed-onset dystonia due to perinatal asphyxia. A prospective study. *Mov Disord* 2007;22(16):2426-9.

ABSTRACT. Dystonias, defined as simultaneous contractions of agonists and antagonists muscles, are expressed in children as abnormal movement or postural changes (dystonic hypertonia). There are pediatric transitional forms (benign and secondary to drugs) and permanent (sequelae, primary or heredodegenerative). The most common forms of dystonia in children is described, highlighting the most prevalent benign dystonia in infant of good performance, dystonia secondary to perinatal noxias (hyperbilirubinemia, asphyxia) or drugs; primary hereditary dystonias and heredodegenerative whose diagnosis allows management and suitable genetic counseling. Among the primary forms of dystonia include early onset DYT-1 and sensitive to dopa and between heredodegenerative disorders glutaric aciduria I and Leigh syndrome are highlighted. The thorough understanding of diverse forms of dystonia in children on the one hand allow direct studies correctly in severe cases to establish prognosis and management and also avoid unnecessary and costly studies in mild and benign pictures.

Keywords: Children, Classification, Dystonia, Muscular dystonia.