



INDIAN JOURNAL OF PRACTICAL PEDIATRICS



- **IJPP is a quarterly subscription journal of the Indian Academy of Pediatrics committed to presenting practical pediatric issues and management updates in a simple and clear manner**
- **Indexed in Excerpta Medica, CABI Publishing, Scopus**

Vol.18 No.2

APR.- JUN. 2016

Dr.P.Ramachandran
Editor-in-Chief

Dr.S.Thangavelu
Executive Editor

CONTENTS

TOPIC OF INTEREST - "PEDIATRIC NEUROLOGY"

Approach to a child with acute flaccid paralysis	101
- Naveen Sankhyan, Renu Suthar	
Approach to a child with ataxia	109
- Leema Pauline C, Viveha Saravanan R, Ravi LA	
Pediatric CNS demyelinating disorders - An update	122
- Lokesh Lingappa, Nikit Milind Shah,	
Approach to muscle disorders in childhood	136
- Viswanathan V	
Hydrocephalus	144
- Hari VS, Thiagarajan G, Lakshmi Tilak S	
Epileptic encephalopathies in children	151
- Vinayan KP	
Neurometabolic disorders: A diagnostic approach	158
- Bindu PS, Arun B Taly	
Traumatic brain injury	171
- Soonu Udani	
Hypoxic ischemic encephalopathy in children: An intensivist's perspective	180
- Jayashree M, Abhijit Choudhary	
Childhood migraine	186
- Sangeetha Yoganathan	

GENERAL ARTICLE

Unexpected difficult pediatric airway: Pearls and pitfalls for the ED physician **193**

- Debasis Das Adhikari, Ekta Rai

DRUG PROFILE

Antacids and H2 antagonists **200**

- Jeeson C Unni, Ranjit Baby Joseph

DERMATOLOGY

Nutritional dermatosis in children **204**

- Anandan V, Yoganandhini C

RADIOLOGY

The acutely swollen limb **210**

- Vijayalakshmi G, Natarajan B

CASE REPORT

Fetal cholelithiasis - A follow up **212**

- Subha B, Parvathy M, Vindyarani WK

ELECTION NOTICE **215,218**

ADVERTISEMENTS **108,192**

CLIPPINGS **121,143,170**

NEWS AND NOTES **121,150,179,185,203,209,211**

BOOK REVIEW **135,199**

FOR YOUR KIND ATTENTION

- * The views expressed by the authors do not necessarily reflect those of the sponsor or publisher. Although every care has been taken to ensure technical accuracy, no responsibility is accepted for errors or omissions.
- * The claims of the manufacturers and efficacy of the products advertised in the journal are the responsibility of the advertiser. The journal does not own any responsibility for the guarantee of the products advertised.
- * Part or whole of the material published in this issue may be reproduced with the note "Acknowledgement" to "Indian Journal of Practical Pediatrics" without prior permission.

- Editorial Board

Published by Dr. P.Ramachandran, Editor-in-Chief, IJPP, on behalf of Indian Academy of Pediatrics, from 1A, Block II, Krsna Apartments, 50, Halls Road, Egmore, Chennai - 600 008. Tamil Nadu, India and printed by Mr. D.Ramanathan, at Alamu Printing Works, 9, Iyyah Street, Royapettah, Chennai-14.

PEDIATRIC NEUROLOGY

APPROACH TO A CHILD WITH ACUTE FLACCID PARALYSIS

***Naveen Sankhyan**
****Renu Suthar**

Abstract: *Acute flaccid paralysis is a clinical syndrome characterized by rapidly evolving weakness, which may include respiratory and bulbar muscles. Acute flaccid paralysis represents a syndromic diagnosis and can have an array of diagnostic possibilities. This condition can be a medical emergency characterized by rapid progress of clinical signs and symptoms. Immediate management includes supporting airway, breathing, and circulation in these children. Diagnosis is clinical and confirmed by specific investigations. An accurate and early etiological diagnosis has an important bearing on the management and prognosis. This review discusses the approach to a child with acute flaccid paralysis and also discusses some key features of the common causes of acute flaccid paralysis.*

Keywords: *Flaccid weakness, Quadripareisis, Polio, Hypotonic paralysis, Guillain-Barre Syndrome.*

Points to Remember

- *Acute flaccid paralysis in children is a medical emergency.*
- *AFP is a clinical syndrome with array of differential diagnosis.*
- *The common causes of AFP are Guillain-Barre syndrome, anterior horn cell myelitis and acute transverse myelitis.*
- *Rapid evolution of the weakness can lead to respiratory failure. Hence a child with AFP should be managed in PICU in the initial few days.*
- *AFP surveillance is a key strategy for global polio eradication.*

References

1. Marx A, Glass JD, Sutter RW. Differential diagnosis of acute flaccid paralysis and its role in poliomyelitis surveillance. *Epidemiol Rev* 2000;22:298-316.
2. Singhi SC, Sankhyan N, Shah R, Singhi P. Approach to a child with acute flaccid paralysis. *Indian J Pediatr* 2012;79:1351-1357.
3. Greninger AL, Naccache SN, Messacar K, Clayton A, Yu G, Somasekar S, et al. A novel outbreak enterovirus D68 strain associated with acute flaccid myelitis cases in the USA (2012-14): a retrospective cohort study. *Lancet Infect Dis* 2015;15:671-682.
4. [www.who.int/immunization/monitoring_surveillance_burden/vpd/surveillance_type/active_poliomyelitis_standards/en/](http://www.who.int/immunization/monitoring_surveillance/burden/vpd/surveillance_type/active_poliomyelitis_standards/en/) accessed 15th march 2016.
5. Bahl S, Kumar R, Menabde N, Thapa A, McFarland J, Swezy V, et al. Polio-free certification and lessons learned-South-East Asia region, March 2014. *MMWR Morb Mortal Wkly Rep* 2014;63:941-946.
6. Messacar K, Abzug MJ, Dominguez SR. 2014 outbreak of enterovirus D68 in North America. *J Med Virol* 2016;88:739-745.
7. Messacar K, Schreiner TL, Maloney JA, Wallace A, Ludke J, Oberste MS, et al. A cluster of acute flaccid paralysis and cranial nerve dysfunction temporally associated with an outbreak of enterovirus D68 in children in Colorado, USA. *Lancet* 2015;385:1662-1671.

* Associate Professor,
Pediatric Neurology and Neurodevelopment Unit,
email: drnsankhyan@yahoo.co.in

** Assistant Professor,
Pediatric Neurology and Neurodevelopment Unit,
Department of Pediatrics,
Postgraduate Institute of Medical Education and Research,
Chandigarh.

8. Hu Y, Jiang L, Peng HL. Clinical Analysis of 134 Children with Nervous System Damage Caused by Enterovirus 71 Infection. *Pediatr Infect Dis J* 2015;34:718-723.
9. van den Berg B, Walgaard C, Drenthen J, Fokke C, Jacobs BC, van Doorn PA. Guillain-Barre syndrome: pathogenesis, diagnosis, treatment and prognosis. *Nat Rev Neurol* 2014;10:469-482.
10. Kuwabara S, Yuki N. Axonal Guillain-Barre syndrome: concepts and controversies. *Lancet Neurol* 2013;12:1180-1188.
11. Sankhyan N, Sharma S, Konanki R, Gulati S. Childhood Guillain-Barre syndrome subtypes in northern India. *J ClinNeurosci* 201;21:427-430.
12. Hughes RA, Swan AV, Raphael JC, Annane D, van Koningsveld R, van Doorn PA. Immunotherapy for Guillain-Barre syndrome: a systematic review *Brain* 2007;130:2245-2257.
13. Melnick J. Enteroviruses: polioviruses, coxsackieviruses, echoviruses, and newer enteroviruses. In: Fields BN, Knipe DM, Chanock RM, eds. *Fields' virology*. 3rd edn. Philadelphia, PA:Lippincott-Raven Publishers, 1996; pp655-712.
14. Teoh HL, Mohammad SS, Britton PN, Kandula T, Lorentzos MS, Booy R, et al. Clinical Characteristics and Functional Motor Outcomes of Enterovirus 71 Neurological Disease in Children. *JAMA Neurol* 2016;19:1-8.
15. Maramattom BV, Philips G, Sudheesh N, Arunkumar G. Acute flaccid paralysis due to West Nile virus infection in adults: A paradigm shift entity. *Ann Indian Acad Neurol* 2014.17:85-88.
16. Holm-Hansen CC, Midgley SE, Fischer TK. Global emergence of enterovirus D68: a systematic review. *Lancet Infect Dis* 2016 Feb 23.pii: S1473-3099(15)00543-5. doi: 10.1016/S1473-3099(15)00543-5. [Epub ahead of print].
17. Transverse Myelitis Consortium Working G. Proposed diagnostic criteria and nosology of acute transverse myelitis. *Neurology* 2002;59:499-505.
18. Suthar R, Sankhyan N, Sahu JK, Khandelwal NK, Singhi S, Singhi P. Acute transverse myelitis in childhood: A single centre experience from North India. *Eur J Paediatr Neurol*. 2016; 20:352-360.
19. Piradov MA, Pirogov VN, Popova LM, Avdunina IA. Diphtheritic polyneuropathy: clinical analysis of severe forms. *Arch Neurol* 2001;58:1438-1442.

PEDIATRIC NEUROLOGY

APPROACH TO A CHILD WITH ATAXIA

***Leema Pauline C**
****Viveka Saravanan R**
****Ravi LA**

Abstract: Ataxia is a relatively common neurological problem in children which encompasses a wide range of causes from infections to inherited disorders. The history and clinical examination coupled with appropriate investigations including neuroimaging can lead to an appropriate diagnosis. Some of these diseases are treatable while the other inherited ataxias require genetic counseling for the patients and their families.

Keywords: Ataxia, Cerebellar, Childhood

Points to remember

- *Ataxias in children may be a manifestation of wide range of disorders.*
- *Diagnosis should be approached with a chronological order into acute, intermittent, and chronic ataxia.*
- *Acute cerebellar ataxia is the most common cause of childhood ataxia which usually results from drug ingestion or postinfectious cerebellar demyelination.*
- *Intermittent ataxia should raise the suspicion of an underlying inborn error of metabolism.*
- *Friedrichs ataxia followed by ataxia telangiectasia are the common causes of inherited ataxias in children.*

References

1. Nejad PJ, Maricich SM, Zoghbi HY. The cerebellum and the hereditary ataxias. In: Swaiman KF, Ashwal S, Ferriero DM, SchorNF, eds. Swaimans Pediatric Neurology, 5th edn Elsevier 2012, pp 939-964.
2. Sivasamy L. Approach to acute ataxia in childhood: Diagnosis and evaluation. *Pediatr Ann* 2014; 43: 153-159.
3. Holmes G. The cerebellum of man. *Brain* 1939;62:1-30.
4. Fogel BL. Childhood Cerebellar Ataxia. *J Child Neurol* 2012;27:1138- 1145.
5. Ryan MM, Engle EC. Acute Ataxia in Childhood. *J Child Neurol* 2003; 18: 309-316.
6. Gosalakkal JA. Ataxias of childhood. *The Neurologist* 2001;7:300-306.
7. Fogel BL, Perlman S. Cerebellar disorders. Balancing the approach to cerebellar ataxia. In: Galvez-Jimenez N, TuiteP, eds. *Uncommon causes of Movement Disorders*, 1st edn. Cambridge University Press New York, 2011; pp 198 -216.
8. Jan MM. Evaluating a child with unsteady gait. *Neurosciences* 2009; 14:3-9.
9. Winchester S, Mikati MA. Ataxia. In: Dulac O, Lassande M, Samat HB, eds. *Handbook of Clinical Neurology*, Amsterdam, Netherlands. Elsevier; 2013: 1213-1217.
10. Mac Donald G P. Ataxia of childhood. In: Berg Bo. Ed. *Child Neurology:A clinical manual*. 2nd edn, JB Lippincott

* Professor of Pediatric Neurology,
email: leemapauline@rediffmail.com

** Assistant Professor of Pediatric Neurology,
Institute of Child Health and Hospital for Children,
Madras Medical College,
Chennai.

& co, Philadelphia, PA 1994; 287-305.

11. Poretti A, Wolf NI, Boltshauser E. Differential diagnosis of cerebellar atrophy in childhood. *Eur J Paediatr Neurol* 2008;12:155-167.
12. Manto M, Marmolino D. Cerebellar ataxias. *Curr Opin Neurol* 2009; 22: 419-429.
13. Lavin MF, Gueven N, Bottle S, Gatti RA. Current and potential therapeutic strategies for the treatment of ataxia telangiectasia. *Br Med Bull* 2007; 81-82: 129-147.
14. Aicardi J, Barbosa C, Andermann E, Morcas R, Ghanem Q, Fukuyama Y, et al. Ataxia oculomotor apraxia: a syndrome mimicking ataxia telangiectasia. *Ann Neurol* 1988;24:497-502.
15. Fogel BL, Perlman S. Clinical features and molecular genetics of autosomal recessive cerebellar ataxias. *Lancet Neurol* 2007; 6: 245-257.
16. Vedolin L, Gonzalez G, Souza CF, Barkovich AJ. Inherited cerebellar ataxia in childhood: A pattern - recognition approach using brain MRI. *Am J Neuroradiol* 2013; 34: 925-934.
17. Jan MM. Neurological examination of difficult and poorly cooperative children. *J Child Neurol* 2007;22: 1209-213.

PEDIATRIC NEUROLOGY

PEDIATRIC CNS DEMYELINATING DISORDERS- AN UPDATE

***Lokesh Lingappa**

****Nikit Milind Shah**

Abstract: Pediatric central nervous system (CNS) demyelinating disorders are a heterogeneous group of conditions with demyelination as the pathological hall mark, acute demyelinating encephalomyelitis (ADEM) being the commonest disorder with very good long term outcomes. Optic neuritis has variable outcome despite aggressive treatment. Transverse myelitis is the most severe of all the demyelinating disorders due to the long term disability it produces. Multiple sclerosis is well known to have a relapsing or progressive course.

Keywords: Demyelinating disorders, Central nervous system, Children.

Points to Remember

- *ADEM is associated with significant involvement of deep grey matter.*
- *Periaqueductal involvement in presence of myelitis or ADEM like picture points towards diagnoses of NMO spectrum disorder.*
- *Second line immunomodulation are increasingly playing a major role in reducing the disability in these demyelinating disorder.*
- *Bilateral optic neuritis is a common association with ADEM whereas unilateral optic neuritis is common as first clinical event of multiple sclerosis.*
- *Multiple sclerosis generally presents as relapsing remitting demyelinating illness.*

References

1. Young NP, Weinshenker BG, Lucchinetti CF. Acute disseminated encephalomyelitis: Current understanding and controversies. *Semin Neurol* 2008; 28(1):84-94 .
2. Leake JA, Albani S, Kao AS, Senac MO, Billman GF, Nespeca MP, et al. Acute disseminated encephalomyelitis in childhood: Epidemiologic, clinical and laboratory features. *Pediatr Infect Dis J* 2004; 23(8):756-764.
3. Marchioni E, Tavazzi E, Minoli L, Del Bue S, Ferrante P, Piccolo G, et al. Acute disseminated encephalomyelitis. *Neurol Sci* 2008;29 (Suppl 2): S286-288.
4. Krupp LB, Banwell B, Tenenbaum S. International Pediatric MS Study Group. Consensus definitions proposed for pediatric multiple sclerosis and related disorders. *Neurology* 2007; 68 (16 Suppl. 2): S7-12.
5. Menge T, Hemmer B, Nessler S, Wiendl H, Neuhaus O, Hartung HP, et al. Acute Disseminated Encephalomyelitis: An Update. *Arch Neurol* 2005;62(11):1673-1680.
6. Pittock SJ1, McClelland RL, Achenbach SJ, Konig F, Bitsch A, Brück W, et al. Clinical course, pathological correlations, and outcome of biopsy proved inflammatory demyelinating disease. *J Neurol Neurosurg Psychiatry* 2005;76(12):1693-1697.
7. Rezai MS, Taghipour M, Azizi F, Abbaskhanian A. Acute Disseminated Encephalomyelitis: A case series and review of literatures. *J Pediatr Rev.* 2013;1:88-98.
8. Garg RK. Acute disseminated encephalomyelitis. *Postgrad Med J* 2003; 79(927):11-17.

* Consultant Child and Adolescent Neurologist

** Consultant Pediatric Neurologist,
Department of Pediatric Neurology and Allied Specialities,
Rainbow Children's Hospital,
Hyderabad.
email : drlokesh@rainbowhospitals.in

9. Ketelslegers IA. Acquired Demyelinating Syndromes and Pediatric Multiple Sclerosis. repub.eur.nl/pub/50369/140122 2014.
10. Mizuguchi M, Abe J, Mikkaichi K, Noma S, Yoshida K, Yamanaka T, et al., Acute necrotising encephalopathy of childhood: a new syndrome presenting with multifocal, symmetric brain lesions. *J Neurol Neurosurg Psychiatry* 1995; 58(5):555-561.
11. Wu X, Wu W, Pan W, Wu L, Liu K, Zhang HL. Acute Necrotizing Encephalopathy: An underrecognized clinicoradiologic disorder. *Mediators Inflamm* 2015;2015:792578. doi: 10.1155/2015/792578.
12. Pena JA, Lotze TE. Pediatric Multiple Sclerosis: Current concepts and consensus definitions. *Autoimmune Dis* 2013;2013:673947. doi: 10.1155/2013/673947
13. Dale RC, Pillai SC. Early relapse risk after a first CNS inflammatory demyelination episode: Examining international consensus definitions. *Dev Med Child Neurol* 2007 Dec;49(12):887-893.
14. Polman CH, Reingold SC, Banwell B, Clanet M, Cohen JA, Filippi M, et al. Diagnostic criteria for multiple sclerosis: 2010 Revisions to the McDonald Criteria. *Ann Neurol* 2011;69(2):292–302.
15. Banwell B, Ghezzi A, Bar-Or A, Mikaeloff Y, Tardieu M. Multiple sclerosis in children: Clinical diagnosis, therapeutic strategies, and future directions. *Lancet Neurol* 2007; 6(10): 887–902.
16. Wingerchuk DM, Banwell B, Bennett JL, Cabre P, Carroll W, Chitnis T, et al. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. *Neurology* 2015 Jul 14;85(2):177-189. doi: 10.1212/WNL.0000000000001729.
17. Nosadini M, Alper G, Riney CJ, Benson LA, Mohammad SS, Ramanathan S, et al. Rituximab monitoring and redosing in pediatric neuromyelitis optica spectrum disorder. *Neurol Neuroimmunol Neuroinflamm* 2016; 3(1): e188. doi: 10.1212/NXI.0000000000000188.

PEDIATRIC NEUROLOGY

APPROACH TO MUSCLE DISORDERS IN CHILDHOOD

***Viswanathan V**

Abstract: *Muscle disorders form a major bulk of cases in any pediatric neurology clinic. It can be quite a daunting task at times to decide on the nature of the muscle disease as there are so many types and the clinical variability is huge. A unique nature of muscle disorders is that they show wide phenotypic variability even in children with the same disease. Two children in the same age group with similar genetic abnormality but showing wide variability in the phenotypic expression of the disease is not uncommon in clinical practice which makes counseling of the family members a difficult task.*

Keywords: *Muscle disorders, Children, Clinical variability, Genetics, Phenotypic expressions, Counseling.*

Points to Remember

- *It is important to make a correct diagnosis of muscle disorders and plan the investigation of choice accordingly.*
- *Genetic diagnosis is helpful in many cases, not only to prognosticate but also to test for carrier status and counsel regarding the chance of recurrence.*
- *Early intervention including physiotherapy, care of respiratory and cardiac issues helps in improving the quality of life and longevity in these children.*
- *Some of the children with muscle disorders may improve over time.*

References

1. Huret JL, Delabar JM, Marlhens F, Aurians A, Berthire M, Tanzer J, et al. 'Down syndrome with duplication of a region of chromosome 21 containing the CuZn superoxide desmutase gene without detectable Karyotypic abnormality. *Human Genetics* 1987;75: 251-257.
2. Munsat TL. Workshop Report: International SMA collaboration. *Neuromuscul Disord* 1991; 1: 81.
3. Vasanth A, Gourie-Devi M, Das S, Gayathri, Ram MY, Anisya V. Neuromuscular disorders in infancy and childhood. *Neurol India* 1997; 45: 63-68.
4. Dubowitz V, *Muscle disorders in childhood*, 2nd Edn, WB Saunders, London, 1995; pp328-329.
5. Spark S, Quijano-Roy S, Harper A, Rutkowski A, Gordon E, Hoffman EP, et al. Congenital muscular dystrophy overview. In: Pagon RA, Adam MP, Ardinger HH, Wallace SE, Amemiya A, Bean LJH, et al, eds. *Gene Reviews* (internet). Seattle (WA): University of Washington, Seattle; 1993-2016. 2001 Jan 22 (updated 2012 Aug 23). <http://www.ncbi.nlm.nih.gov/books/NBK1291/>
6. Philpot J, Cowan F, Pennock J, Sewry C, Dubowitz V, Bydder G, et al. Merosin deficient muscular dystrophy: the spectrum of brain involvement on magnetic resonance imaging. *Neuromuscul Disord* 1999; 9: 81-85.
7. Saito K, Fukuyama Congenital Muscular Dystrophy (Jan 26th 2006; last updated May 10th 2012). In: Pagon RA, Adam MP, Ardinger HH, Wallace SE, Amemiya A, Bean LJH, et al, eds. *Gene Reviews* (internet) Seattle (WA); University of Washington, Seattle 1993 -2014.

* Consultant Pediatric Neurologist,
Kanchi Kamakoti CHILDS Trust and
Apollo Children's Hospitals,
Chennai.
email : vishneuro@gmail.com

8. Butler MG, Meaney FJ, Palmer CG. Clinical and cytogenetic survey of 39 individuals with Prader-Labhart Willi Syndrome Am J Med Genet 1986;23: 793-809.
9. Knoll HM, Nichols RD, Magenis RE, Graham DM, Lalonde M, Latt SA. Angelman and Prader-Willi syndromes share a common chromosome 15q deletion but differ in parental origin of the deletion. Am J Med Genet 1989;32:285-290.
10. Lefvert AK, Osterman PO. Newborn infants born to myasthenic mothers : a clinical study and an investigation of acetyl choline receptor antibodies in 17 children. Neurology 1983;33:133-138.
11. Hers HG. Alpha Glucosidase deficiency in generalized glycogen storage disease (Pompe's disease) Biochem J 1963;86:11-16.
12. Katzin LW, Amato AA. Pompe disease: a review of the current diagnosis and treatment recommendations in the era of enzyme replacement therapy. J Clin Neuromuscul Dis 2008;9: 421-431.
13. Hoffman EP, Fischbeck KH, Brown RH, Johnson M, Medori R, Loike JD, et al. Characterization of dystrophin in muscle biopsy specimens from patients with Duchenne or Becker muscular dystrophy. N Eng J Med 1988;318: 1363-1368.
14. Hoffman EP, Kunkel LM, Angelini C, Clarke A, Johnson M, Harris JB. Improved diagnosis of Becker Muscular Dystrophy by dystrophin testing. Neurology 1989;39: 1011-1017.
15. Nalini A, Gayathri N, Thaha F, Das S, Shylashree S. Sarcoglycanopathies - Clinical and histochemical characteristics in 66 patients. Neurol India 2010; 58(5):691-696.
16. Abicht A, Müller J S, Lochmüller H. Congenital Myasthenic Syndromes. 2003 May 9 [Updated 2012 Jun 28]. Pagon RA, Adam MP, Ardinger HH, Wallace SE, Amemiya A, Bean LJH , et al., eds. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2014. Available from: <http://www.ncbi.nlm.nih.gov/books/NBK1168/>.

PEDIATRIC NEUROLOGY

HYDROCEPHALUS

***Hari VS**
 ****Thiagarajan G**
 *****Lakshmi Tilak S**

Abstract: Pathophysiology, clinical features, management and complications of hydrocephalus have been discussed in this article. Emphasis is on practical aspects relevant to the pediatrician and residents. Procedures have been dealt with in detail wherever necessary. Endoscopic third ventriculostomy which has become safer and improvements in shunt systems are discussed.

Keywords: Hydrocephalus, Childhood, Shunt, CSF.

Points to Remember

- *Serial assessment of head circumference charting and matching it with age and sex matched graphs to identify hydrocephalus and early shunting in case of clinical signs and symptoms of raised ICP is advised.*
- *Cognitive improvement is better with early shunting.*

References

1. Aschoff A, Kremer P, Hashemi B, Kunze S. The scientific history of hydrocephalus and its treatment. *Neurosurg Rev* 1999;22:67-93.
2. Kulkarni AV, Drake JM, Kestle JR, Mallucci CL, Sgouros S, Constantini S. Endoscopic Third Ventriculostomy Vs Cerebrospinal fluid shunt in the treatment of Hydrocephalus: A Propensity Score- Adjusted Analysis. *J Neurosurgery*. 2010; 67(3):588-593.
3. Abou-Hamden A, Drake JM. Hydrocephalus. In: Albright L, Pollack I, Adelson D, eds. Principles and Practice of Paediatric Neurosurgery, 3rd edn. New York: Thieme Medical Publishers, 2015;pp89-97.
4. Greenberg MS. Hydrocephalus. In: Handbook of Neurosurgery, 6th Edn, Greenberg Graphics, Lakeland, FL, 2006;pp180-199.
5. Chazal J. Management of Hydrocephalus in Childhood, In: Marc Sindou, ed. Practical Handbook of Neurosurgery, 1st Edn, Springer Wien New York, 2009; pp525-536.
6. Chidambaram B. CSF diversionary procedures. In: Ravi R, Sridhar K, Vasudevan M, eds. Textbook of Operative Neurosurgery, 1st edition, New Delhi, BI Publications Pvt Ltd, 2005; pp291-301.
7. Scott RM. Shunt Complications. In: Rangachary SS, Wilkins RH, eds. 2nd edn, Neurosurgery. New York: McGraw-Hill;1996: pp 3655-3664.
8. Cohen AR, Perneckzy A. Endoscopy and the management of third ventricular lesions. In: Apuzzo MLJ (ed) Surgery of the third ventricle, 2nd edn. Williams and Wilkins, Baltimore, USA 1996; pp889-936.
9. Walker ML, Fried A, Petronio J. Diagnosis and treatment of the slit ventricle syndrome. *Neurosurg Clin N Am* 1993; 4:707-714.
10. Sciubba DM, Stuart RM, McGirt MJ, Woodworth GF, Samdani A, Carson B, et al. Effect of antibiotic-impregnated shunt catheters in decreasing the incidence of shunt infection in the treatment of hydrocephalus. *J Neurosurg* 2005, 103(Suppl 2):131-136.

* Associate Professor
email : vshari@yahoo.com

** Professor

*** Assistant Professor,
Department of Neurosurgery,
Pondicherry Institute of Medical Sciences, Pondicherry.

PEDIATRIC NEUROLOGY

EPILEPTIC ENCEPHALOPATHIES IN CHILDREN

***Vinayan KP**

Abstract: *Childhood epileptic encephalopathic syndromes are a group of conditions in which cognitive, sensory and/or motor functions deteriorate as a consequence of epileptic activity. This terminology classically denotes a group of well-defined epileptic syndromes of childhood associated with a high probability of encephalopathic features that develop or worsen after the onset of epilepsy. However, it is increasingly being used in children who develop any deterioration/stagnation in development as a result of presumed epileptic activity. This phenomenon is most common and severe in infancy and early childhood. Evidence for the currently available therapeutic options in these difficult-to-treat epileptic syndromes is reviewed and a stepwise management strategy is suggested.*

Keywords: *Epileptic encephalopathy, Syndrome, Children, Treatment*

Points to remember

- *Childhood epileptic encephalopathic syndromes are a group of resistant epilepsies in which cognitive, sensory, and/or motor functions deteriorate as a consequence of epileptic activity.*
- *This deterioration may be the product of the underlying cause, the result of epileptic activity in the brain, or a combination of both.*
- *The currently available therapeutic options in epileptic encephalopathies include antiepileptic drugs, co-factors like pyridoxine, steroids, intravenous immunoglobulins, epilepsy surgery and ketogenic diet.*
- *Children with epileptic encephalopathic syndromes may need early and aggressive management in a specialized center for the optimal developmental outcome.*

References

1. Nabbout R, Dulac O. Epileptic Encephalopathies: A Brief Overview. *J Clin Neurophysiol* 2003;20:393–397.
2. Berg AT, Berkovic SF, Brodie MJ, Buchhalter J, Cross JH, van Emde Boas W, et al. Revised terminology and concepts for organization of seizures and epilepsies: report of the ILAE Commission on Classification and Terminology, 2005-2009. *Epilepsia* 2010; 51:676-685.
3. Wirrell E, Farrell K, Whiting S. The epileptic encephalopathies of infancy and childhood. *Canadian J Neurol Sci* 2005; 32: 409-418.
4. Besag FM. Cognitive and behavioral outcomes of epileptic syndromes: implications for education and clinical practice. *Epilepsia* 2006;47(suppl.2): 119-125.
5. Holmes GL, Ben-Ari Y. The neurobiology and consequences of epilepsy in the developing brain. *Pediatr Res* 2001; 49:320-325.
6. Vinayan KP. Epilepsy, antiepileptic drugs and educational problems. *Indian Pediatr* 2006; 43:786-794.
7. Schubert R. Attention deficit disorders and epilepsy. *Pediatr Neurol* 2005; 32: 32:1-10.
8. Loring DW, Meador KJ. Cognitive side effects of antiepileptic drugs in children. *Neurology* 2004; 62: 872-877.

* Professor and Head,
Division of Pediatric Neurology,
Department of Neurology
Amrita Institute of Medical Sciences,
Cochin.
e-mail: vinayankp@aims.amrita.edu

9. Farwell JR, Lee YJ, Hirtz DG, Sulzbacher SI, Ellenberg JH, Nelson KB. Phenobarbital for febrile seizures-effects on intelligence and on seizure recurrence. *New Engl J Med* 1990; 322:364-369.
10. Darke K, Edwards SW, Hancock E, Johnson AL, Kennedy CR, Lux AL, et al. Developmental and epilepsy outcomes at age 4 years in the UKISS trial comparing hormonal treatments to vigabatrin for infantile spasms: A multi-centre randomised trial. *Arch Dis Child* 2010; 95:382-386.
11. Lux AL, Edwards SW, Hancock E, Johnson AL, Kennedy CR, Newton RW, et al. The United Kingdom infantile spasms study (UKISS) comparing hormone treatment with vigabatrin on developmental and epilepsy outcomes to age 14 months: A multicentre randomised trial. *Lancet Neurol* 2005;4:712-717.
12. Roulet-Perez E, Davidoff V, Mayor-Dubois C, Maeder-Ingvar M, Seeck M, Ruffieux C, et al. Impact of severe epilepsy on development: Recovery potential after successful early epilepsy surgery. *Epilepsia* 2010;51:1266-1276.
13. Elterman RD, Shields WD, Mansfield KA, Nakagawa J. Randomized trial of vigabatrin in patients with infantile spasms. *Neurology* 2001;57:1416-1421.
14. Hancock EC, Osborne JP, Edwards SW. Treatment of infantile spasms. *Cochrane Database of Systematic Reviews* 4:2008; CD 001770.
15. Loddenkemper T, Fernandez IS, Peters JM. Continuous spike and waves during sleep and electrical status epilepticus in sleep. *J Clin Neurophysiol* 2011;28:154-164.
16. Van Lierde A. Therapeutic data. In: Beaumanoir A, Bureau M, Deonna T, MiraL, Tassinari CA (Eds). *Continuous Spikes and Waves During Slow Sleep, Electrical Status Epilepticus During Slow Sleep: Acquired Epileptic Aphasia and Related Conditions*. John Libbey, London 1995; pp225-227.
17. De Negri M, Baglietto MG, Battaglia FM, Gaggero R, Pessagno A, Recanati L. Treatment of electrical status epilepticus by short diazepam (dzp) cycles after dzp rectal bolus test. *Brain Dev* 1995;17: 330-333.
18. Chiron C, Marchand MC, Tran A, Rey E, d'Athis P, Vincent J, et al. Stiripentol in severe myoclonic epilepsy in infancy: A randomised placebo-controlled syndrome-dedicated trial. *Sticlo study group. Lancet* 2000;356:1638-1642.
19. Inoue Y, Ohtsuka Y, Oguni H, Tohyama J, Baba H, Fukushima K, et al. Stiripentol open study in Japanese patients with Dravet syndrome. *Epilepsia* 2009;50:2362-2368.
20. Corda D, Gelisse P, Genton P, Dravet C, Baldy-Moulinier M. Incidence of drug-induced aggravation in benign epilepsy with centrotemporal spikes. *Epilepsia* 2001;42:754-759.
21. Chiron C, Dulac O. The pharmacologic treatment of Dravet syndrome. *Epilepsia* 2011;52 (Suppl 2):72-75.
22. Sakakihara Y. Treatment of West syndrome. *Brain Dev* 2011;33:202-206.
23. Lux AL, Edwards SW, Hancock E, Johnson AL, Kennedy CR, Newton RW, et al. The United Kingdom infantile spasms study comparing vigabatrin with prednisolone or tetracosactide at 14 days: A multicentre, randomised controlled trial. *Lancet* 2004;364:1773-1778.
24. Pellock JM, Hrachovy R, Shinnar S, Baram TZ, Bettis D, Dlugos DJ, et al. Infantile spasms: A U.S. Consensus report. *Epilepsia* 2010;51:2175-2189.
25. Mikati MA, Kurdi R, El-Khoury Z, Rahi A, Raad W. Intravenous immunoglobulin therapy in intractable childhood epilepsy: Open-label study and review of the literature. *Epilepsy Behav* 2010;17:90-94.
26. Specchio N, Fusco L, Claps D, Vigeveno F. Epileptic encephalopathy in children possibly related to immune mediated pathogenesis. *Brain Dev* 2010;32:51-56.
27. Freitag H, Tuxhorn I. Cognitive function in preschool children after epilepsy surgery: rationale for early intervention. *Epilepsia* 2005;46:561-567.
28. Freeman JM, Kossoff EH, Hartman AL. The ketogenic diet: one decade later. *Pediatrics* 2007;119:535-543.
29. Caraballo RH. Nonpharmacologic treatments of Dravet Syndrome: Focus on the ketogenic diet. *Epilepsia* 2011;52(Suppl. 2):79-82.
30. Caraballo RH, Cersosimo RO, Saks D, Cresta A, Escobal N, Fejerman N. Ketogenic diet in patients with myoclonic astatic epilepsy. *Epileptic Disord* 2006;8:151-155.

PEDIATRIC NEUROLOGY

NEUROMETABOLIC DISORDERS: A DIAGNOSTIC APPROACH

***Bindu PS**

****Taly AB**

Abstract: *Inherited neurometabolic disorders constitute an important group of genetic disorders with diverse neurological manifestations. Many of them are amenable for treatment and early intervention is necessary to prevent or ameliorate the extent of brain damage. However, the diagnosis and management of these disorders are often challenging to the clinicians in view of the overlapping and non-specific phenotypes. A systematic diagnostic approach often helps in narrowing down the differential diagnoses and plan appropriate investigations. This review presents a symptom-based approach for diagnosis of common metabolic disorders encountered in clinical practice.*

Keywords: *Neurometabolic disorders, Inborn errors of metabolism, Epilepsy, Children.*

Points to Remember

- *Neurometabolic disorders cause diverse neurological manifestations.*
- *A systematic approach encompassing clinical, biochemical and magnetic resonance imaging helps in diagnosis.*
- *It is important to be familiar with the age-dependent manifestations of the common neurometabolic disorders.*
- *The rational treatment of metabolic disorders requires understanding of the pathophysiological process responsible for the disease.*
- *Early intervention can improve the quality of life and prevent irreversible brain damage.*

References

1. Cassis L, Cortes-Saladelafont E, Molero-Luis M, Delia Yubero, Maria Julieta González, Aida Ormazabal Herrero, et al. Review and evaluation of the methodological quality of the existing guidelines and recommendations for inherited neurometabolic disorders. *Orphanet J Rare Dis* 2015;10:164.
2. Gropman AL. Patterns of brain injury in inborn errors of metabolism. *Semin Pediatr Neurol* 2012; 19: 203-210.
3. van Karnebeek CD, Shevell M, Zschocke J, Moeschler JB, Stockler S. The metabolic evaluation of the child with an intellectual developmental disorder: diagnostic algorithm for identification of treatable causes and new digital resource. *Mol Genet Metab.* 2014; 111: 428-438.
4. van Karnebeek CD, Stockler S. Treatable inborn errors of metabolism causing intellectual disability: a systematic literature review. *Mol Genet Metab.* 2012; 105: 368-381.
5. Saudubray JM and Charpentier C. Clinical phenotypes: Diagnosis/Algorithms. In: Scriver CR BA, Valle D, Sly WS, Childs B, Kinzler KW, et al, editors, (ed.). *The Metabolic and Molecular Basis of Inherited Disease* 8th edn. New York: McGraw-Hill 2001; pp1327-403.
6. Maegawa GH, Stockley T, Tropak M, Banwell B, Blaser S, Kok F, et al. The natural history of juvenile or subacute GM2 gangliosidosis: 21 new cases and literature review of 134 previously reported. *Pediatrics* 2006; 118: e1550-1562.

* Additional Professor
email : drpsbindu@yahoo.co.in

** Professor,
Department of Neurology,
National Institute of Mental Health and Neurosciences,
Bangalore.

7. Shapiro BE, Logigian EL, Kolodny, Pastores GM. Late-onset Tay-Sachs disease: the spectrum of peripheral neuropathy in 30 affected patients. *Muscle Nerve* 2008; 38: 1012-1015.
8. Zaroff CM, Neudorfer O, Morrison C, Pastores GM, Rubin H, Kolodny EH. Neuropsychological assessment of patients with late onset GM2 gangliosidosis. *Neurology* 2004; 62: 2283-2286.
9. Duffner PK, Barczykowski A, Jalal K, Yan L, Kay DM, Carter RL. Early infantile Krabbe disease: results of the World-Wide Krabbe Registry. *Pediatr Neurol* 2011; 45: 141-148.
10. Duffner PK, Barczykowski A, Kay DM, Jalal K, Yan L, Abdelhalim A, et al. Later onset phenotypes of Krabbe disease: results of the world-wide registry. *Pediatr Neurol* 2012; 46: 298-306.
11. Abdelhalim AN, Alberico RA, Barczykowski AL, Duffner PK. Patterns of magnetic resonance imaging abnormalities in symptomatic patients with Krabbe disease correspond to phenotype. *Pediatr Neurol* 2014; 50: 127-134.
12. Saudubray JM, Ogier H, Bonnefont JP, Munnich A, Lombes A, Hervé F, et al. Clinical approach to inherited metabolic diseases in the neonatal period: a 20-year survey. *J Inherit Metab Dis* 1989; 12 Suppl 1: 25-41.
13. Alston CL, He L, Morris AA, Hughes I, de Goede C, Turnbull DM, et al. Maternally inherited mitochondrial DNA disease in consanguineous families. *Eur J Hum Genet* 2011; 19: 1226-1229.
14. Clarke JTR. *A Clinical Guide to Inherited Metabolic Diseases*. 3rd edn, Cambridge University Press, New York, 2005.
15. Lee RW, Poretti A, Cohen JS, Levey E, Gwynn H, Johnston MV, et al. A diagnostic approach for cerebral palsy in the genomic era. *Neuromolecular Med* 2014; 16: 821-844.
16. Renaud DL. Leukoencephalopathies associated with macrocephaly. *Semin Neurol* 2012; 32: 34-41.
17. Poll-The BT, Maillette de Buy Wenniger-Prick LJ, Barth PG, Duran M. The eye as a window to inborn errors of metabolism. *J Inherit Metab Dis* 2003; 26: 229-244.
18. Nagappa M, Bindu PS, Taly AB, Sinha S and Bharath RD. *Child Neurology: Molybdenum cofactor deficiency*. *Neurology* 2015; 85: e175-178.
19. Bindu PS, Christopher R, Mahadevan A and Bharath RD. Clinical and imaging observations in isolated sulfite oxidase deficiency. *J Child Neurol* 2011; 26: 1036-1040.
20. Wakabayashi K, Gustafson AM, Sidransky E and Goldin E. Mucopolipidosis type IV: an update. *Mol Genet Metab* 2011; 104: 206-213.
21. Bindu PS, Gayathri N, Yasha TC, et al. A variant form of mucopolipidosis IV: report on 4 patients from the Indian subcontinent. *J Child Neurol* 2008; 23: 1443-1446.
22. Wolf NI, Garcia-Cazorla A, Hoffmann GF. Epilepsy and inborn errors of metabolism in children. *J Inherit Metab Dis* 2009; 32: 609-617.
23. Sedel F, Fontaine B, Saudubray JM and Lyon-Caen O. Hereditary spastic paraparesis in adults associated with inborn errors of metabolism: a diagnostic approach. *J Inherit Metab Dis* 2007; 30: 855-864.
24. Bindu PS, Mahadevan A, Taly AB, Christopher R, Gayathri N, Shankar SK. Peripheral neuropathy in metachromatic leucodystrophy. A study of 40 cases from south India. *J Neurol Neurosurg Psychiatry* 2005; 76: 1698-1701.
25. Siddiqi ZA, Sanders DB, Massey JM. Peripheral neuropathy in Krabbe disease: electrodiagnostic findings. *Neurology* 2006; 67: 263-267.
26. Garcia-Cazorla A, Wolf NI, Serrano M, Perez-Duenas B, Pineda M, Campistol J, et al. Inborn errors of metabolism and motor disturbances in children. *J Inherit Metab Dis* 2009; 32: 618-29.
27. Debray FG, Lambert M, Gagne R, Maranda B, Laframboise R, MacKay N, et al. Pyruvate dehydrogenase deficiency presenting as intermittent isolated acute ataxia. *Neuropediatrics* 2008; 39: 20-23.
28. Wang D, Pascual JM, Yang H, Engelstad K, Jung S, Sun RP, et al. Glut-1 deficiency syndrome: clinical, genetic, and therapeutic aspects. *Ann Neurol* 2005; 57: 111-118.
29. Christopher R, Sankaran BP. An insight into the biochemistry of inborn errors of metabolism for a clinical neurologist. *Ann Indian Acad Neurol* 2008; 11: 68-81.
30. Schiffmann R, van der Knaap MS. Invited article: an MRI-based approach to the diagnosis of white matter disorders. *Neurology* 2009; 72: 750-759.
31. Leach EL, Shevell M, Bowden K, Sylvia Stockler-Ipsiroglu, Clara DM van Karnebeek. Treatable inborn errors of metabolism presenting as cerebral palsy mimics: systematic literature review. *Orphanet J Rare Dis* 2014; 9:197.

PEDIATRIC NEUROLOGY

TRAUMATIC BRAIN INJURY

***Soonu Udani**

Abstract: Traumatic brain injury (TBI) is the most common cause of intracranial hypertension. The hallmark of TBI is cerebral edema and raised intracranial pressure with its detrimental effect on the brain. The focus here is on the practical aspects of controlling intracranial pressure, maintaining cerebral perfusion pressure and supporting the patient's hemodynamics and vital functions during the initial critical days.

Keywords: Traumatic brain injury, Intracranial pressure, Cerebral perfusion pressure, Neuromonitoring.

Points to Remember

- *Basic monitoring and meticulous care is of prime importance in the management of traumatic brain injury.*
- *Seizures, fever, pain and sedation need close attention.*
- *CPP targeted therapies hold promise for better outcomes.*
- *ICP monitoring is useful for targeting therapy.*
- *Hypertonic saline is preferred over mannitol.*
- *Hypothermia is advised only in refractory life threatening ICP.*
- *Attention not only to mortality but also to good outcome is vital.*

References

1. Gururaj G. Epidemiology of traumatic brain injuries: Indian scenario. *Neurol Res* 2002 24(1):24-28.
2. Werner C, Engelhard K. Pathophysiology of traumatic brain injury. *Br J Anaesth* 2007; 99 :4-9.
3. Head injury: assessment and early management. NICE guidelines [CG176] Published date: January 2014
4. Bramwell KJ, Haizlip J, Pribble C, VanDerHeyden, TC, Witte M. The effect of etomidate on intracranial pressure and systemic blood pressure in pediatric patients with severe traumatic brain injury. *Pediatr Emerg Care*. 2006; 22:90-93.
5. Brussel T, Thiessen J, Vigfusson G, Lunkenheimer P, Aken H, Lawin P. Hemodynamic and cardiodynamic effects of propofol and etomidate: negative inotropic properties of propofol. *Anesth Analg*. 1989; 69:35-40.
6. Lewandowski-Belfer JJ, Patel AV, Darracott RM, Jackson DA, Nordeen JD. Safety and Efficacy of Repeated Doses of 14.6 or 23.4 % Hypertonic Saline for Refractory Intracranial Hypertension. *Neurocrit Care* 2014; 20(3): 436-42. doi: 10.1007/s12028-013-9907-1
7. Gonda DD, Meltzer HS, Crawford JR, Hilfiker ML, Shellington DK, Peterson BM, Levy ML. Complications Associated With Prolonged Hypertonic Saline Therapy in Children With Elevated Intracranial Pressure. *Ped Crit Care Med* 2013; 14(6):601-620.

* Pediatric Intensivist,
Section Head - Pediatrics,
PD Hinduja Hospital, Mumbai.
email : drsudani@gmail.com

8. Skippen P, Seear M, Poskitt K, Kestle J, Cochrane D, Annich G, et al. Effect of hyperventilation on regional cerebral blood flow in head-injured children. *Crit Care Med*. 1997; 25:1402–1409.
9. Cochran A, Scaife ER, Hansen KW, Downey EC. Hyperglycemia and outcomes from pediatric traumatic brain injury. *J Trauma*. 2003; 55:1035–1038.
10. Inaba K, Menaker J, Branco BC, Gooch J, Okoye OT, Herrold J. A prospective multicenter comparison of levetiracetam versus phenytoin for early posttraumatic seizure prophylaxis. *J Trauma Acute Care Surg* 2013;74: 766–773.
11. Jones KE, Puccio AM, Harshman KJ, Falcione J, Benedict B, Jankowitz BT. Levetiracetam versus phenytoin for seizure prophylaxis in severe traumatic brain injury. *Neurosurg Focus*,25(4), 2008E3.
12. Szaflarski JP. Is There Equipose Between Phenytoin and Levetiracetam for Seizure Prevention in Traumatic Brain Injury? *Epilepsy Curr*. 2015; 15(2): 94–97.
13. Kochanek PM, Carney N, Adelson PD, Ashwal S, Bell MJ, Bratton S, et al. Guidelines for the acute medical management of severe traumatic brain injury in infants, children, and adolescents—second edition. *Pediatr Crit Care Med*. 2012; 13 (Suppl 1):S1–S82.
14. Allen BB, Chiu YL, Gerber LM, Ghajar J, Greenfield JP. Age-specific cerebral perfusion pressure thresholds and survival in children and adolescents with severe traumatic brain injury *Pediatr Crit Care Med*. 2014; 15:62–70.
15. Udani S. Advances in neurocritical care; *Indian J Pediatr*. 2015 ;82(3):272-276.
16. Kurtz P, Gaspard N, Wahl AS, Bauer RM, Hirsch LJ, Wunsch H, et al. Continuous electroencephalography in a surgical intensive care unit. *Int Care Med* 2014 ;40(2):228-234.
17. Bourdages M, Bigras JL, Farrell CA, Hutchison JS, Lacroix J. Cardiac arrhythmias associated with severe traumatic brain injury and hypothermia therapy. *Pediatr Crit Care Med* 2010; 11(3): 408-11
18. Adelson PD, Wisniewski SR, Beca J, Brown SD, Bell M, Muizelaar JP, et al. Comparison of hypothermia and normothermia after severe traumatic brain injury in children (Cool Kids):a phase 3, randomised controlled trial. *Lancet Neurol* 2013; 12(6):546-553.
19. Mayer SA, Chong J. Critical care management of increased intracranial pressure. *J Intensive Care Med* 2002;17:55-67.
20. Fearnside MR, Cook RJ, McDougall P, McNeil RJ. The Westmead Head Injury Project outcome in severe head injury. A comparative analysis of pre-hospital, clinical and CT variables. *Br J Neurosurg*. 1993;7(3):267-279.
21. Beers SR, Wisniewski SR, Garcia-Filion P, Tian Y, Hahner T, Berger RP, et al. Validity of a pediatric version of the Glasgow Outcome Scale-Extended. *J Neurotrauma*. 2012;29(6): 1126-1139.
22. Anderson VA, Catroppa C, Haritou F, Morse S, Pentland L, Rosenfeld J, et al. Predictors of Acute Child and Family Outcome following Traumatic Brain Injury in Children. *Pediatr Neurosurg* 2001;34:138–148.
23. Wilde EA, Hunter JV, Newsome MR, Scheibel RS, Bigler ED, Johnson JL. Frontal and Temporal Morphometric Findings on MRI in Children after Moderate to Severe Traumatic Brain Injury. *Journal of Neurotrauma*. 2005, 22(3): 333-344.

PEDIATRIC NEUROLOGY

HYPOXIC ISCHEMIC ENCEPHALOPATHY IN CHILDREN: AN INTENSIVIST'S PERSPECTIVE

***Jayashree M**
****Abhijit Choudhary**

Abstract: Hypoxic ischemic encephalopathy is a syndrome of acute global neuronal injury resulting from combination of hypoxia, ischemia and reperfusion. The clinical conditions leading to HIE in children include cardiac arrest, asphyxia and drowning. Anatomical areas of the brain vulnerable to hypoxia and ischemia include hippocampus, caudate and putamen with relative sparing of the brainstem. The most common clinical presentation is altered consciousness. The key objective of intensive care management is to anticipate, prevent and treat secondary physiological insults to the brain through a structured protocolized 'neuroprotective approach'. Therapeutic hypothermia is coming up as an option for older children especially in out-of-hospital cardiac arrest once return of spontaneous circulation is achieved. Outcome goals are quality survival rather than survival alone.

Keywords: Hypoxic ischemic encephalopathy, Children, Intensive care, Neuroprotection.

Points to Remember

- *Hypoxic-ischemic encephalopathy is a constellation of pathophysiological and molecular injuries induced by hypoxia, ischemia and cytotoxicity and further aggravated by reperfusion.*
- *Children with HIE present with altered consciousness and seizures.*
- *Intensive care management focuses on maintaining a balance between systemic and cerebral targets with the help of multimodal neuromonitoring.*
- *The neuroprotective approach emphasizes on normoxia, normocarbica, normotension, and euglycemia.*
- *Fever must be strictly avoided in all children with HIE.*
- *Therapeutic hypothermia can be considered as an option in OHCA after ROSC is achieved.*
- *Biomarkers, evoked potentials and neuroimaging have been used in combination with clinical signs for outcome prediction.*

References

1. Busl KM, Greer DM. Hypoxic-ischemic brain injury: pathophysiology, neuropathology and mechanisms. *NeuroRehabilitation* 2010;26(1):5–13.
2. Harukuni I, Bhardwaj A. Mechanisms of brain injury after global cerebral ischemia. *Neurol Clin* 2006;24(1):1–21.
3. Nadkarni VM, Larkin GL, Peberdy MA, Carey SM, Kaye W, Mancini ME, et al. First documented rhythm and clinical outcome from in-hospital cardiac arrest among children and adults. *JAMA* 2006; 295(1):50–57.
4. Rathore V, Bansal A, Singhi SC, Singhi P, Muralidharan J. Survival and neurological outcome following in-hospital paediatric cardiopulmonary resuscitation in North India. *Paediatr Int Child Health* 2016; 36(2):141-147.
5. Manole MD, Foley LM, Hitchens TK, Kochanek PM, Hickey RW, Bayir H, et al. Magnetic resonance imaging assessment of regional cerebral blood flow after asphyxial cardiac arrest in immature rats. *J Cereb Blood Flow Metab* 2009; 29(1):197–205.
6. Back T, Hemmen T, Schüler OG. Lesion evolution in cerebral ischemia. *J Neurol* 2004; 251(4):388–397.

* Professor and Incharge, Pediatric Emergency and ICU,
email : mjshree@hotmail.com

** Senior Resident, Pediatric Critical Care
Advanced Pediatrics Centre,
PGIMER, Chandigarh.

7. MacPherson HA, Mackinaw-Koons B, Leffler JM, Fristad MA. Pilot Effectiveness Evaluation of Community-Based Multi-Family Psychoeducational Psychotherapy for Childhood Mood Disorders. *Couple Fam Psychol* 2016; 5(1):43–59.
8. Ramesh Kumar R, Singhi SC, Singhi P. Raised intracranial pressure (ICP): management in emergency department. *Indian J Pediatr* 2012;79(4):518–524.
9. Bohn DJ, Biggar WD, Smith CR, Conn AW, Barker GA. Influence of hypothermia, barbiturate therapy, and intracranial pressure monitoring on morbidity and mortality after near-drowning. *Crit Care Med* 1986;14(6):529–534.
10. Le Roux PD, Jardine DS, Kanev PM, Loeser JD. Pediatric intracranial pressure monitoring in hypoxic and nonhypoxic brain injury. *Childs Nerv Syst ChNS Off J Int Soc Pediatr Neurosurg* 1991;7(1):34–39.
11. Rittenberger JC, Popescu A, Brenner RP, Guyette FX, Callaway CW. Frequency and timing of nonconvulsive status epilepticus in comatose post-cardiac arrest subjects treated with hypothermia. *Neurocrit Care* 2012; 16(1):114–122.
12. Reynolds JC, Lawner BJ. Management of the post-cardiac arrest syndrome. *J Emerg Med* 2012; 42(4):440–449.
13. Gluckman PD, Wyatt JS, Azzopardi D, Ballard R, Edwards AD, Ferriero DM, et al. Selective head cooling with mild systemic hypothermia after neonatal encephalopathy: multicentre randomised trial. *Lancet Lond Engl* 2005; 365(9460):663–670.
14. Shankaran S, Laptook AR, Ehrenkranz RA, Tyson JE, McDonald SA, Donovan EF, et al. Whole-body hypothermia for neonates with hypoxic-ischemic encephalopathy. *N Engl J Med* 2005; 353(15):1574–1584.
15. Kleinman ME, Chameides L, Schexnayder SM, Samson RA, Hazinski MF, Atkins DL, et al. Part 14: pediatric advanced life support: 2010 American Heart Association Guidelines for Cardiopulmonary Resuscitation and Emergency Cardiovascular Care. *Circulation* 2010; 122(18 Suppl 3):S876–908.
16. Young KD, Gausche-Hill M, McClung CD, Lewis RJ. A prospective, population-based study of the epidemiology and outcome of out-of-hospital pediatric cardiopulmonary arrest. *Pediatrics* 2004; 114(1):157–164.
17. Slonim AD, Patel KM, Ruttimann UE, Pollack MM. Cardiopulmonary resuscitation in pediatric intensive care units. *Crit Care Med* 1997; 25(12):1951–1955.
18. Booth CM, Boone RH, Tomlinson G, Detsky AS. Is this patient dead, vegetative, or severely neurologically impaired? Assessing outcome for comatose survivors of cardiac arrest. *JAMA* 2004; 291(7):870–879.
19. Carter BG, Butt W. A prospective study of outcome predictors after severe brain injury in children. *Intensive Care Med* 2005; 31(6):840–845.
20. Mandel R, Martinot A, Delepoulle F, Lamblin M-D, Laureau E, Vallee L, et al. Prediction of outcome after hypoxic-ischemic encephalopathy: a prospective clinical and electrophysiologic study. *J Pediatr* 2002; 141(1):45–50.
21. Tiainen M, Roine RO, Pettilä V, Takkunen O. Serum neuron-specific enolase and S-100B protein in cardiac arrest patients treated with hypothermia. *Stroke J Cereb Circ* 2003; 34(12):2881–2886.
22. Pfeifer R, Börner A, Krack A, Sigusch HH, Surber R, Figulla HR. Outcome after cardiac arrest: predictive values and limitations of the neuroproteins neuron-specific enolase and protein S-100 and the Glasgow Coma Scale. *Resuscitation* 2005; 65(1):49–55.
23. Fink EL, Manole MD, Clark RS. Hypoxic-Ischemic Encephalopathy. In: Rogers 'Textbook of pediatric intensive care, 5th edn, Lippinkott Williams & Wilkins, Woulter Kluwer, 2015;p1043.

PEDIATRIC NEUROLOGY

CHILDHOOD MIGRAINE

***Sangeetha Yoganathan**

Abstract: Headache is one of the most common causes of referral to the emergency department and neurology clinic visit in children. Migraine, tension type headache and cluster headache are the common causes of primary headache. Based on the temporal patterns, headache can be categorized into acute, acute and recurrent, chronic non-progressive and chronic and progressive. The pathophysiology of migraine is complex. The traditional medical model of history, general physical and neurological examination should be followed in evaluation of any child with headache. General physical examination begins with assessment of vitals, anthropometry and search for any external markers of vasculitis. Thorough neurological examination should be carried out to document any signs of raised intracranial pressure or focal neurological deficits. Imaging is not routinely recommended in children with well recognized episodic headache symptoms suggesting diagnosis of migraine. Nonsteroidal anti-inflammatory agents (NSAIDs), acetaminophen, 5-HT receptor agonists, dopamine receptor antagonists, and antihistamine agents are often used in aborting acute migraine attacks. Migraine headache that impairs the quality of life and functioning are indicators for the initiation of prophylaxis. This review will briefly discuss about the clinical approach, evaluation, differential diagnosis and management of children with migraine.

Keywords: Headache, Migraine, Migraine prophylaxis

Points to Remember

- *Migraine is a common cause of headache in children and adolescents.*
- *Migraine without aura is the most frequent form.*
- *Diagnosis is essentially clinical and other causes of headache such as neoplastic, vascular, metabolic or toxic disorders must be excluded.*
- *Need for prophylaxis is decided by the headache burden and disability.*
- *Balanced treatment with pharmacological measures and biobehavioural interventions should be endorsed.*

References

1. Al-Twajiri WA, Shevell MI. Pediatric migraine equivalents: Occurrence and clinical features in practice. *Pediatr Neurol* 2002; 26:365-368.
2. Deubner DC. An epidemiologic study of migraine and headache in 10-20 year olds. *Headache* 1977; 17:173-180.
3. Sillanpaa M. Changes in the prevalence of migraine and other headache during the first seven school years. *Headache* 1983; 23:15-19.
4. Mortimer MJ, Kay J, Jaron A. Epidemiology of headache and childhood migraine in an urban general practice using Ad Hoc, Vahlquist and IHS criteria. *Dev Med Child Neurol* 1992; 34:1095-1101.
5. Rasmussen BK, Olesen J. Migraine with aura and migraine without aura: An epidemiological study. *Cephalalgia* 1992; 12:221-228.
6. Rothner AD. Headaches in children: A review. *Headache* 1978; 18:169-175.
7. Headache Classification Committee of the International Headache Society (IHS). The International Classification of Headache Disorders, 3rd edn beta version. *Cephalalgia* 2013; 33:629-808.
8. Kruuse C, Thomsen LL, Birk, S, Olesen J. Migraine can be induced by sildenafil without changes in middle cerebral artery diameter. *Brain* 2003; 126:241-247.
9. Pietrobon D, Striessnig J. Neurobiology of migraine. *Nat Rev Neurosci* 2003; 4:386-398.
10. Rothner AD. The evaluation of headaches in children and adolescents. *Semin Pediatr Neurol* 1995; 2:109-118.
11. Conicella E, Raucci U, Vanacore N, Vigevano F, Reale A,

* Associate Professor,
Department of Neurological Sciences,
Christian Medical College,
Vellore.
email : doc_ys@yahoo.co.in

- Pirozzi N, et al. The child with headache in a pediatric emergency department. *Headache* 2008; 48:1005-1011.
12. Scagni P, Pagliero R. Headache in an Italian pediatric emergency department. *J Headache Pain* 2008; 9:83-87.
 13. The epidemiology of headache among children with brain tumor. Headache in children with brain tumors. The Childhood Brain Tumor Consortium. *J Neurooncol* 1991; 10:31-46.
 14. Lewis DW, Qureshi F. Acute headache in children and adolescents presenting to the emergency department. *Headache* 2000; 40:200-203.
 15. Callaghan BC, Kerber KA, Pace RJ, Skolarus L, Cooper W, Burke JF. Headache neuroimaging: Routine testing when guidelines recommend against them. *Cephalalgia* 2015; 35:1144-1152.
 16. Lewis D, Ashwal S, Hershey A, Hirtz D, Yonker M, Silberstein S. American Academy of Neurology Quality Standards Subcommittee; Practice Committee of the Child Neurology Society. Practice parameter: pharmacological treatment of migraine headache in children and adolescents: report of the American Academy of Neurology Quality Standards Subcommittee and the Practice Committee of the Child Neurology Society. *Neurology*. 2004;63:2215-2224.
 17. Arora S, Wagner JG, Herbert M. Myth: parenteral ketorolac provides more effective analgesia than oral ibuprofen. *Can J Emerg Med Care* 2007; 9:30-32.
 18. Loga P, Lewis D. Compazine in Migraine. *Emergency Medicine Journal (BMJ)*. 2007:297-298.
 19. Sheridan DC, Spiro DM, Meckler GD. Pediatric migraine: abortive management in the emergency department. *Headache* 2014; 54:235-245.
 20. Bachur RG, Monuteaux MC, Neuman MI. Comparison of acute treatment regimens for migraine in the emergency department. *Pediatrics* 2015; 135:232-238.
 21. Gelfand AA, Goadsby PJ. Treatment of pediatric migraine in the emergency room. *Pediatr Neurol* 2012; 47:233-241.
 22. Lewis DW. Pediatric migraine. *Neurol Clin* 2009;27:481-501.
 23. Sheridan DC, Spiro DM, Nguyen T, Koch TK, Meckler GD. Low dose propofol for the abortive treatment of pediatric migraine in the emergency department. *Pediatr Emerg Care* 2012; 28:1293-1296.
 24. Hickman C, Lewis KS, Little R, Rastogi RG, Yonker M. Prevention for Pediatric and Adolescent Migraine. *Headache* 2015; 55:1371-1381.
 25. Schetzek S, Heinen F, Kruse S, Borggraefe I, Bonfert M, Gaul C, et al. Headache in children: update on complementary treatments. *Neuropediatrics* 2013; 44: 25-33.
 26. Condò M, Posar A, Arbizzani A, Parmeggiani A. Riboflavin prophylaxis in pediatric and adolescent migraine. *J Headache Pain* 2009; 10:361-365.
 27. MacLennan SC, Wade FM, Forrest KM, Ratanayake PD, Fagan E, Antony J. High-dose riboflavin for migraine prophylaxis in children: a double-blind, randomized, placebo-controlled trial. *J Child Neurol* 2008; 23: 1300-1304.
 28. Bruijn J, Duivenvoorden H, Passchier J, Locher H, Dijkstra N, Arts WF. Medium-dose riboflavin as a prophylactic agent in children with migraine: a preliminary placebo-controlled, randomised, double-blind, crossover trial. *Cephalalgia* 2010; 30:1426-1434.
 29. Di Lorenzo C, Pierelli F, Coppola G, Grieco GS, Rengo C, Ciccolella M, et al. Mitochondrial DNA haplogroups influence the therapeutic response to riboflavin in migraineurs. *Neurology* 2009; 72: 1588-1594.
 30. Tepper SJ. Nutraceutical and Other Modalities for the Treatment of Headache. *Continuum (Minneapolis)* 2015; 21:1018-1031.
 31. Lewis DW. Migraine and migraine variants in childhood and adolescence. *Semin Pediatr Neurol* 1995; 2:127-143.
 32. Kropp P, Meyer B, Landgraf M, Ruscheweyh R, Ebinger F, Straube A. Headache in children: update on biobehavioral treatments. *Neuropediatrics* 2013; 44:20-24.
 33. Sun-Edelstein C, Mauskop A. Alternative headache treatments: nutraceuticals, behavioral and physical treatments. *Headache* 2011; 51:469-483.
 34. Guyuron B, Varghai A, Michelow B, Thomas T, Davis J. Corrugator supercilii muscle resection and migraine headaches. *Plast Reconstr Surg* 2000; 106:429-434.
 35. Binder WJ, Brin MF, Blitzer A, Schoenrock LD, Pogoda JM. Botulinum toxin type A (BOTOX) for treatment of migraine headaches: an open-label study. *Otolaryngol Head Neck Surg* 2000; 123:669-676.
 36. Guyuron B. Is Migraine Surgery Ready for Prime Time? The Surgical Team's View. *Headache* 2015; 55:1464-1473.
 37. Monastero R, Camarda C, Pipia C, Camarda R. Prognosis of migraine headaches in adolescents: a 10-year follow-up study. *Neurology* 2006; 67:1353-1356.

GENERAL ARTICLE

UNEXPECTED DIFFICULT PEDIATRIC AIRWAY: PEARLS AND PITFALLS FOR THE EMERGENCY DEPARTMENT PHYSICIAN

***Debasis Das Adhikari**

****Ekta Rai**

Abstract: *Unexpected difficult pediatric airway without predictors is rare but when encountered is a nightmare. These crises can be salvaged safely most of the time if the background knowledge, concepts and strategies are not only read but also rehearsed, practiced and discussed frequently. As the pediatric emergency doctors rarely face the problems of maintaining unanticipated difficult airway, they have to be well versed with the guidelines. This review article proposes to present the simple stepwise approach to such a situation based on the current evidences and literature.*

Keywords: *Difficult airway, Unanticipated, Children, Emergency.*

Points to Remember

- *Be familiar and be prepared with alternative methods of intubating techniques and use it regularly in your day-to-day practice as part of failure plans, e.g. laryngeal mask airway, gum elastic bougie, fiber optic intubation so that you will not fumble at the time of crisis and will not panic.*
- *Oxygenate at all times as oxygenation is more important than intubation in the time of crisis.*
- *“It is preferable to use superior judgment – to avoid having to use superior skill.”*
- *Step-by-step process is in order.*
- *Help should be called for early.*

References

1. Sims C, von Ungern-Sternberg BS. The normal and the challenging pediatric airway; Paediatr Anaesth 2012; 22: 521–526.
2. Weiss M, Engelhardt T. Proposal for the management of the unexpected difficult pediatric airway. Paediatr Anaesth 2010; 20: 454–464.
3. Holm-Knudsen RJ, Rasmussen LS. Pediatric airway management: Basic aspects. Acta Anaesthesiol Scand 2009; 53: 1-9.
4. Medina AR, Gómez LR, Ospina OA, Ocampo F. The pediatric airway: concepts to bear in mind during anesthetic management. Rev Colomb Anestesiología 2012; 40(3):199-202.
5. Caplan RA, Benumof JL, Berry FA, Blitt CD, Bode RH, Cheney FW et al. Practice guidelines for management of difficult airway. A report by the American Society of Anesthesiologists Task Force on management of difficult airway. Anesthesiology 1993; 78:597-602.
6. Benumof JL. The American Society of Anesthesiologists' management of the difficult airway algorithm and explanation: Analysis of the algorithm. In Benumof JL (ed): Airway Management Principles and Practice. 1997; St. Louis, Mosby-Year Book.
7. Gupta S, Sharma R, Jain D. Airway assessment: predictors of difficult airway. Indian J Anaesth 2005; 49(4):257-262.

* Pediatric Emergency Medicine

** Department of Anaesthesia,
Department of Pediatrics,
Christian Medical College, Vellore.
email : debasis@cmcvellore.ac.in

8. Heinrich S, Birkholz T, Ihmsen H, Irouschek A, Ackermann A, Schmidt J. Incidence and predictors of difficult laryngoscopy in 11,219 pediatric anesthesia procedures. *Paediatr Anaesth* 2012; 22:729–36.
9. Auroy Y, Ecoffey C, Messiah A. Relationship between complications of pediatric anesthesia and volume of pediatric anesthetics. *Anesth Analg* 1997; 84: 234–235.
10. Lerman J, Creighton RE. Two hands, three sites: show me the vocal cords. *Pediatr Anaesth* 2006; 16: 96.
11. Kheterpal S, Martin L, Shanks AM, Tremper KK. Prediction and outcomes of impossible mask ventilation: a review of 50,000 anesthetics. *Anesthesiology* 2009; 110: 891–897.
12. Wheeler M. Management strategies for the difficult pediatric airway. *Anesthesiol Clin North Am* 1998; 16: 743–761.
13. Black AE, Flynn PER, Smith HL, Thomas ML, Wilkinson KA. Development of guideline for the management of the unanticipated difficult airway in pediatric practice. *Paediatr Anaesth* 2015; 25: 346-362.
14. Knill RL. Difficult laryngoscopy made easy with a BURP. *Can J Anaesth* 1993; 40: 279-82.
15. Henderson JJ, Popat MT, Latto IP, Pearce AC. Difficult Airway Society guidelines for management of the unanticipated difficult intubation. *Anaesthesia* 2004; 59: 675–694.
16. Walker RW. The laryngeal mask airway in the difficult paediatric airway: an assessment of positioning and use in fiberoptic intubation. *Paediatr Anaesth* 2000; 10:53-58.
17. Frova G, Guarino A, Petrini F, Merli G, Sorbellom, SARNePI, et al. Recommendations for airway control and difficult airway management in pediatric patients. *Minerva Anesthesiol* 2006; 72: 723–748.
18. Metterlein T, Frommer M, Kwok P, Lyer S, Graf BM, Sinner B. Emergency cricothyrotomy in infants - Evaluation of a novel device in an animal model. *Pediatric Anaesthesia* 2011; 21: 104-109.

DRUG PROFILE

ANTACIDS AND H₂ RECEPTOR ANTAGONISTS

***Dr. Jeelson C Unni**

****Dr Ranjit Baby Joseph**

Abstract: Strong evidence for the use of antacids and to a lesser extent, Histamine H₂ receptor antagonists in the treatment of conditions requiring a reduction in gastric acid production is lacking. Other indications for specific antacid molecules are also discussed.

Keywords: Antacids, Histamine H₂ receptor blockers, Proton pump inhibitors.

Points to Remember

- *PPIs are the drug of choice in primary acid reflux disease.*
- *Efficacy of H₂RAs is not proved in the treatment of GERD but may be used in conditions associated with acute gastrointestinal bleed.*
- *H₂RAs may reduce severity of the refluxate.*
- *Antacids are not routinely recommended for treatment of GERD. Calcium salts are recommended as phosphate binders and its use as antacids should be avoided.*

References

1. Goodman & Gilman's the pharmacological basis of therapeutics. 12th edn. Eds Laurence L. Brunton, Bruce A Chabner, Bjorn C Knollmann. New York: McGraw-Hill 2011;pp1310-1317.
2. Hahn D, Hodson EM, Craig JC. Interventions for metabolic bone disease in children with chronic kidney disease. Cochrane Database Syst Rev 2015 Nov 12;11:CD008327. doi: 10.1002/14651858.CD008327.pub2.
3. Wang Y, Xie G, Huang Y, Zhang H, Yang B, Mao Z. Calcium acetate or calcium carbonate for hyperphosphatemia of hemodialysis patients: a meta-analysis. PLoS One 2015; 10(3): e0121376.doi:10.1371/journal.pone.0121376.eCollection 2015.
4. IAP Drug Formulary. 4th edn. Eds Jeelson C Unni, Menon PSN, Nair MKC, Bansal CP. 2015, Publication of IAP. Pixel Studio, Cochin, 2015;pp227-228.
5. IAP Drug Formulary. 4th edn. Eds Jeelson C Unni, Menon PSN, Nair MKC, Bansal CP. 2015, Publication of IAP. Pixel Studio, Cochin, 2015;p261.
6. Joint Formulary Committee. British National Formulary for children. London: BMJ Group and Pharmaceutical Press, 2013-2014; p477.
7. van der Pol R, Langendam M, Benninga M, van Wijk M, Tabbers M. Efficacy and safety of histamine-2 receptor antagonists. JAMA Pediatr 2014;168(10):947-54. doi: 10.1001/jamapediatrics.2014.1273.
8. Tighe M, Afzal NA, Bevan A, Hayen A, Munro A, Beattie RM. Pharmacological treatment of children with gastro-oesophageal reflux. Cochrane Database Syst Rev 2014; 11:CD008550. Epub 2014 Nov 24.

* Editor-in-Chief
IAP Drug Formulary,
Associate Consultant in Pediatrics
e-mail : jeelson1955@gmail.com

** Specialist in Pediatrics,
Aster Medcity,
Kochi.

9. Rostom A, Dube C, Wells G, Tugwell P, Welch V, Jolicoeur E, McGowan J. Prevention of NSAID-induced gastroduodenal ulcers. *Cochrane Database Syst Rev* 2002; (4): CD002296.
10. Zhang YS, Li Q, He BS, Liu R, Li ZJ. Proton pump inhibitors therapy vs H2 receptor antagonists therapy for upper gastrointestinal bleeding after endoscopy: A meta-analysis. *World J Gastroenterol* 2015; 21(20): 6341-6351. doi: 10.3748/wjg.v21.i20.6341.
11. IAP Drug Formulary 2015. 4th Ed. Eds Jeesson C Unni, Menon PSN, Nair MKC, Bansal CP. 2015, Publication of IAP. Pixel Studio, Cochin: 480.
12. IAP Drug Formulary 2015. 4th Ed. Eds Jeesson C Unni, Menon PSN, Nair MKC, Bansal CP. 2015, Publication of IAP. Pixel Studio, Cochin: 281.
13. Kirch W, Hoensch H, Janisch HD. Interactions and non-interactions with ranitidine. *Clin Pharmacokinet* 1984;9(6):493-510.
14. IAP Drug Formulary 2015. 4th edn. Eds Jeesson C Unni, Menon PSN, Nair MKC, Bansal CP. 2015, Publication of IAP. Pixel Studio, Cochin, 2015;p337.

DERMATOLOGY**NUTRITIONAL DERMATOSIS IN CHILDREN**

***Anandan V**
****Yoganandhini C**

Abstract: *Skin is the most important organ providing sensory perception, enclosing barrier and environmental protection, regulating temperature and producing vitamin D. Nutrition is a dynamic process concerned with ingestion, digestion, absorption and assimilation of food for nourishing the body. Skin reflects the internal well being and balanced nutrition in the form of smooth shiny skin, glossy hair, well developed muscles, bones and teeth, strong build and energetic to look at.*

Keywords: *Micronutrients, Deficiency status, Macronutrients, Nutritional dermatoses.*

Points to Remember

- *Skin can reflect nutritional deficiency.*
- *Sound knowledge about dermatological manifestations of nutritional deficiency will help in early diagnosis.*
- *Correction of the deficiency will reverse the cutaneous findings.*

References

1. Miller SJ. Nutritional deficiency and the skin. *J Am Acad Dermatol* 1989;21:1-30.
2. Prendiville J, Manfredi L. Skin signs of nutritional disorders. *Semin Dermatol* 1992; (11)1: 88-97.
3. Thacher TD, Fischer PR, Pettifor JM, Darmstadt GL. Nutritional rickets in ichthyosis and response to calcipotriene. *Pediatrics* 2004;114(1):e119-123.
4. Roe DA. Riboflavin deficiency: Mucocutaneous signs of acute and chronic deficiency. *Semin Dermatol* 1991;10(4):293-295.
5. Cakmak SK, Gönül M, Aslan E, Gül U, Kiliç A, Heper AO. Half-and-half nail in a case of pellagra. *Eur J Dermatol* 2006;16(6): 695-696.
6. Graells J, Ojeda RM, Muniesa C, Gonzalez J, Saavedra J. Glossitis with linear lesions: An early sign of vitamin B12 deficiency. *J Am Acad Dermatol* 2009;60(3):498-500.
7. Rushton DH. Nutritional factors and hair loss. *Clin Exp Dermatol* 2002;27(5):396-404.
8. Traupe H, Happle R, Gröbe H, Bertram HP. Polarisation microscopy of hair in acrodermatitis enteropathica. *Pediatr Dermatol* 1986;3(4): 300-303.

* Prof and HOD

** Junior Resident,

Department of Dermatology,

Stanley Medical College, Chennai.

e-mail : dermanandan@gmail.com

CASE REPORT**FETAL CHOLELITHIASIS – A FOLLOW UP**

***Subha B**
****Parvathy M**
*****Vindyarani WK**

Abstract: *Fetal gallstones, detected by routine third trimester ultrasound, have been described in the literature with controversial clinical significance. We report a case of fetal cholelithiasis detected at 38 weeks gestation during a routine scan. The patient remained asymptomatic and had a complete spontaneous resolution of the gallstones in postnatal life as described in most other studies.*

Keywords: *Fetal cholelithiasis, Follow up, Ultrasonogram*

References

1. Brown DL, Teele RL, Doubilet PM, DiSalvo DN, Benson CB, Van Alstyne GA. Echogenic material in the fetal gallbladder: Sonographic and clinical observations. *Radiology* 1992;182:73-76.
2. Gertner M, Farmer D. Laparoscopic cholecystectomy in a 16-day-old infant with chronic cholelithiasis. *J Pediatr Surg* 2004;39:17-19.
3. Stringer MD, Lim P, Cave M, Martinez D, Liford RJ. Fetal gallstones. *J Pediatr Surg* 1996;31:1589-1591.
4. Iroh Tam PY, Angelides A. Perinatal detection of gallstones in siblings. *Am J Perinatol* 2010;27:771-774.
5. Abbitt PL, McIlhenny J. Prenatal detection of gallstones. *J Clin Ultrasound* 1990;18:202-204.
6. Sheiner E, Abramowicz JS, Hershkovitz R. Fetal gallstones detected by routine third trimester ultrasound. *Int J Gynaecol Obstet* 2006;92:255-256.
7. Cancho Candela R, Díaz González J, Perandones Fernández C, Viñuela Rueda B, Relea Sarabia A, Andrés de Llano JM. Echogenic material in fetal gallbladder: prenatal diagnosis and postnatal follow-up [in Spanish] *An Pediatr (Barc)* 2004;61:326-9
8. Fanaroff AA, Martin RJ, Miler MJ. Identification and management of high-risk problems in the neonate. In: Creasy RK, Resnik R, eds. *Maternal-Fetal Medicine, Principles and Practice*. 2nd edn. Philadelphia, PA: Saunders 1989; pp 1176-1179.
9. Suchet IB, Labatte MF, Dyck CS, Salgado LA. Fetal cholelithiasis: A case report and review of the literature. *J Clin Ultrasound* 1993;21:198-202
10. Munjuluri N, Elgharaby N, Acolet D, Kadir RA. Fetal gallstones. *Fetal Diagn Ther* 2005;20:241-243.

* Assistant Professor

** Associate Professor

*** Professor and Head,

Department of Pediatrics,

Sri Muthukumaran Medical College and Research Institute,

Chennai.

e-mail : subhavinish@gmail.com