Therapeutic Uses of IVIG in Children — Ramesh and Schwartz

Cerebral Palsy — Taft

Management of Fatal Illness and Death in Children or Their Parents — Barakat, Sills, LaBagnara
CONTENTS

ARTICLES

403 Therapeutic Uses of Intravenous Immunoglobulin (IVIG) in Children
Sujatha Ramesh and Stanley A. Schwartz

411 Cerebral Palsy
Lawrence T. Taft

419 Management of Fatal Illness and Death in Children or Their Parents
Lamia P. Barakat, Richard Sills, Susan LaBagnara

426 Consultation with the Specialist: Degenerative Central Nervous System Disease
Andrew G. Vaducz and Leon G. Epstein

433 Index of Suspicion
John Kidd, Donald L. Batisky, Constantine A. Stratakis, Adolpho Garnica, Benjamin R. Waller III, Landon B. Pendergrass

DEPARTMENT OF CORRECTIONS

436 Erratum

IN BRIEF

424 Cleft Palate
432 Rubella Vaccine
437 Muscular Dystrophy
438 Antibiotic Drug Levels

COVER

Working in the medium of batik, Paul Nzalamba creates images that are drawn from his native country, Uganda, and that reflect the strength, struggle, and beauty of all people, especially children and adolescents. We chose to use his “At Play” (1988) to show a modern, indigenous artist’s work that illustrates the color and joy of such artists. Mr. Nzalamba’s works are on display at his studio in Los Angeles, California. Reproduced with permission.

ANSWER KEY
INFECTIOUS DISEASE
Immunoglobulin Therapy

SUGGESTED READING
Dwyer JM. Thirty years of supplying the missing link. Am J Med. 1984;76:46–52
Mazer BD, Gelfand EW. An open-label study of high-dose intravenous immunoglobulin in severe childhood asthma. J Allergy Clin Immunol. 1991;87:976–983


PIR QUIZ

1. A major advantage of immunoglobulin administered intravenously compared with that administered intramuscularly is:
   A. High doses can be given, with little discomfort to the patient.
   B. It can be administered to patients younger than 6 months of age.
   C. Long-term immunosuppressive effects are avoided.
   D. Patients who have selective IgA deficiency can be treated safely.

2. It is the current consensus that the administration of intravenous immunoglobulin in the neonatal age group is most beneficial for:
   A. Infants born of mothers who have active genital herpes.
   B. Preterm infants who have birthweights less than 2500 g.
   C. Prevention of respiratory syncytial virus infection in high-risk infants.
   D. Seriously ill infants who are septic as an addition to more aggressive therapy.

3. The most correct statement regarding the effect of therapy with IVIG in childhood is:
   A. Acute status asthmaticus is resolved by large doses.
   B. Monthly administration to children who have juvenile rheumatoid arthritis prolongs remissions.
   C. Remission can be induced in idiopathic thrombocytopenic purpura.
   D. The number of episodes of acute otitis media is reduced significantly.

4. A 3-year-old Caucasian girl is admitted with a 1-week history of sore throat and spiking fever to 104°F (40°C). She was seen by her pediatrician 3 days ago. Her throat culture was negative for Streptococcus. Amoxicillin therapy has not resulted in improvement. Immunizations are current. On physical examination she appears toxic but alert. She has bilateral conjunctivitis; an erythematous tongue and pharynx; bilateral, tender anterior cervical lymphadenopathy; edematous hands and feet that have desquamation; and a diffuse, macular erythematous rash over her trunk and proximal extremities. Her white blood cell count is 13600/mm3 with 62% mature neutrophils, 27% nonskewed (band) neutrophils, and 11% mature lymphocytes; her hemoglobin level is 12.5 g/dL; and her platelet count is 800,000/mm3. The tentative diagnosis is Kawasaki disease. The most appropriate therapeutic plan is:
   A. Administer IVIG in a dose of 2 g/kg promptly together with aspirin.
   B. Delay IVIG therapy until the diagnosis is confirmed.
   C. Give IVIG 400 mg/kg for 4 consecutive days.
   D. Start IVIG therapy on first evidence of coronary artery disease.

5. The diagnosis of common variable immunodeficiency has been established in a 6-year-old child. The most correct statement with respect to treating this child with IVIG is:
   A. A negative purified protein derivative test for tuberculosis is a prerequisite to initiating treatment with IVIG.
   B. IVIG should be administered monthly and serum levels monitored.
   C. The preferred program to reduce anaphylaxis is alternate administration of IVIG and intramuscular gamma globulin.
   D. The rate of human immunodeficiency virus in the IVIG product must be balanced against the need for protective antibodies.

6. The adverse effects of IVIG therapy include all of the following except:
   A. Abdominal cramps accompanied by vomiting.
   B. Back pains and leg cramps.
   C. Chest tightness and wheezing.
   D. Chills and fever.
   E. Generalized petechiae and ecchymoses.
tional potential, but the achievement of these expectations also depends on psychosocial adjustment.

Children who have hemiparesis walk by 1.5 to 3 years of age; 80% to 90% of children who have diplegia, 70% of those who have dyskinesia, and 50% who have quadriplegia attain some mode of ambulation. In these clinical types the ability to maintain independent sitting by 1.5 to 2 years offers a good prognosis for community ambulation. Children who learn to sit between 2 and 4 years usually become household ambulators, and some may walk short distances outdoors with assistive devices. Walking is not expected of children who cannot sit by 4 years of age. Despite the unilateral hand impairment, hemiparesis does not exclude independence in daily activities. Motor control of the upper extremities is adequate to perform activities of daily living in diplegia and usually in cases of mild ambulatory quadriplegia. Some children who have quadriplegia and rely on wheelchair mobility may achieve partial independence in self-care. About 25% of this group requires help for all activities. Athetoid and ataxic movement disorders tend to compromise fine hand dexterity to a greater extent than ambulation. Mental retardation delays but does not exclude walking if it is permitted by the motor disability. Achievements in daily living skills are consistent with intellectual competence, regardless of the degree of physical disability.

SUGGESTED READING
Cleft Palate

An Overview of Middle Ear Disease in Cleft Palate Children. Muniz HR. Facial Plast Surg. 1993;9:177-180


Cleft palate (CP) is one of the most common congenital malformations in children. Isolated CP occurs in approximately 1 in 2500 newborns; cleft lip with or without CP occurs in about 1 in 700 live births. Caused by failure of the embryonic palatine shelves to fuse in the midline between the sixth and tenth weeks of gestation, CP actually represents a complex of problems involving not only a defect in the palate, but dysfunction of the eustachian tubes and middle ears as well. Thus, the newborn who has a cleft may have difficulty sucking and feeding and may go on to develop hypernasal speech, a condition that leads to problems with clarity of speech. The child may be subject to recurrent episodes of acute and/or serious otitis media, which themselves may lead to conductive hearing loss. Dental problems may be associated with the cleft, and cleft also may be a marker for an underlying multiple malformation syndrome with numerous additional medical problems. For all of these reasons, early detection of the palatal defect is essential.

Early detection, however, is not always easy. CP actually is a spectrum of disorders, ranging from frank defects in both the soft and hard palate, readily seen on peroral examination, to bifid uvula, which often is missed or considered little more than a normal variant. Between these two extremes lies the occult submucous cleft, a defect in the soft palate covered by mucosa and often difficult to diagnose because it appears closed. A bluish, V-shaped discoloration of the posterior palate may be the only sign that such a defect exists.

An important subgroup of CP is the Pierre Robin malformation sequence (PRMS), consisting of three separate but related anatomic problems: micrognathia (small jaw), glossoptosis (tongue obstructing the airway), and a U-shaped cleft of the soft (and sometimes the hard) palate. A single malformation, failure of the jaw to grow properly during a critical stage early in the first trimester of pregnancy, leads to the entire sequence. Pressed upward into an aberrant position by the small jaw, the tongue, which actually is normal in size, forces the palatine shelves to fuse around it, resulting in a U-shaped palatal defect. Finally, because the oral cavity is small (again, due to the undergrown jaw), the tongue falls back against the airway, leading to symptomatic obstructive apnea.

Obstructive apnea in a newborn who has a U-shaped cleft palate is a medical emergency. The apnea will not be detected by a cardiorespiratory monitor because the chest wall continues to move, but there is no effective air exchange. Unless an oral airway is inserted, the newborn who has PRMS may asphyxiate. A definitive procedure, usually a glossoptomy, rarely a tracheostomy, may be required to assure patency of the airway. Obstructive apnea in children who have PRMS is self-limited; within approximately 1 year their mandibles almost always have grown sufficiently to allow the tongue to fall away from the airway. At that point, the glossoptomy or tracheostomy can be removed and the cleft repaired.

Even infants who have frank CP but not PRMS may develop symptoms during the immediate newborn period. Effective sucking depends on the ability to generate negative pressure; children who have CP and cannot seal the oral cavity because of the palatal defect usually are poor feeders and may fail to thrive. Although opinions differ about breast-
<table>
<thead>
<tr>
<th>ONSET</th>
<th>DISORDER/ETIOLOGY</th>
<th>FEATURES</th>
<th>INITIAL SYMPTOMS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Birth</td>
<td>GM1 gangliosidosis type 1: deficiency of lysosomal beta galactosidase</td>
<td>• Poor suck</td>
<td>WHITE MATTER X</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Hypotonia</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Hepatosplenomegaly</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td>• Coarse features</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Seizures</td>
<td></td>
</tr>
<tr>
<td>1–3 weeks</td>
<td>Maple syrup urine disease: deficiency of branched chain ketoacid dehydrogenase</td>
<td>Well at birth</td>
<td>GRAY MATTER X</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Deterioration following initiation of feeding</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td>• Recurrent emesis</td>
<td></td>
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<td></td>
<td></td>
<td>• Progressive lethargy</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Seizures</td>
<td></td>
</tr>
<tr>
<td>1–6 months</td>
<td>GM2 gangliosidosis type 1: deficiency of hexosaminidase A (Tay-Sachs)</td>
<td>• Psychomotor regression</td>
<td>WHITE MATTER X</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Excessive startle response</td>
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<tr>
<td></td>
<td></td>
<td>• Cherry-red macula</td>
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<td></td>
<td></td>
<td>• Spasticity</td>
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<tr>
<td></td>
<td></td>
<td>• Seizures</td>
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</tr>
<tr>
<td></td>
<td></td>
<td>• Blindness</td>
<td></td>
</tr>
<tr>
<td>1–6 months</td>
<td>Globoid cell leukodystrophy (Krabbe disease): deficiency of galactosylceramide beta-galactosidase</td>
<td>• Hyperirritability</td>
<td>GRAY MATTER X</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Unexplained fever</td>
<td></td>
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<td></td>
<td></td>
<td>• Feeding difficulty</td>
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<td>• Psychomotor regression</td>
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<td>• Spasticity</td>
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<td>• Blindness</td>
<td></td>
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<tr>
<td>1–2 years</td>
<td>Infantile metachromatic leukodystrophy: deficiency of aryl-sulfatase A</td>
<td>• Gait ataxia</td>
<td>GRAY MATTER X</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Hypotonia with hyporeflexia followed by spasticity</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Mental deterioration</td>
<td></td>
</tr>
</tbody>
</table>