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COVER

"Le Gourmet," painted in 1901 by Picasso (1881-1973) during his "Blue Period," demonstrates the natural appetite of the small child, who appears well nourished and even is eating standing up. Eating problems in children are not inherent in their stage of development but are their response to adverse environments. The blue color, however, suggests a threat to this healthy state. Child health professionals must balance this innate healthy aspect of childhood against the environmental threats to their well-being and be advocates for the healthy development of children. (This painting is from the National Gallery of Art's Chester Dale collection and is reproduced with permission.)

ANSWER KEY


Pediatrics in Review
Vol 15 No 9
September 1994

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PEDIATRICS IN REVIEW (ISSN 0191-9601) is owned and controlled by the American Academy of Pediatrics. It is published monthly by the American Academy of Pediatrics, 141 Northwest Point Blvd, PO Box 927, Elk Grove Village, IL 60009-0927. Statements and opinions expressed in Pediatrics in Review are those of the authors and not necessarily those of the American Academy of Pediatrics or its Committees. Recommendations included in this publication do not indicate an exclusive course of treatment or serve as a standard of medical care.

Subscription price for 1994: AAP Fellow $100; AAP Candidate Fellow $75, AAFP $125; Allied Health or Resident $70; Nonmember or Institution $130. Current single price is $10. Subscription changes will be honored up to 12 months from the publication date.

Second-class postage paid at ARLINGTON HEIGHTS, ILLINOIS 60009-0927 and at additional mailing offices.

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The printing and production of Pediatrics in Review is made possible, in part, by an educational grant from Ross Products Division, Abbott Laboratories.
down in the pelvic colon to determine the depth of descent of the rectal pouch accurately. Children generally are studied at this point unless they become distended or demonstrate the presence of a perineal fistula. Such patients should be made NPO and begin intravenous antibiotics. Although NG tubes are not placed routinely, if the child needs to be transferred to a referral center for appropriate diagnosis and management, we believe that the NG tube should be used during transport.

Children who have imperforate anus often will manifest the VATER associations and need to be investigated carefully for other anomalies. A significant feature of this anomaly is the absence of sacral vertebral segments in some children who have high type defects; these children can be predicted to have some deficiencies in their neurologic control of defecation. Almost 50% of children who have high imperforate anus also will have genital anomalies; an ultrasonographic study of the kidney and early assessment of the renal excretory status is important in the preoperative evaluation.

**MANAGEMENT**

In general, most low type imperforate anus problems can be managed via a perineal anoplasty performed during infancy. High imperforate anus, however, requires a more extensive operative repair as well as division of the likely rectourethral fistula. Therefore, these infants generally are managed via a colostomy in the newborn period. In addition to stool diversion, the properly constructed colostomy will avoid urinary tract soiling via contamination of the rectourethral fistula. Definitive reconstructive pull-through surgery then is undertaken at about 8 months of age.

Children who have low type imperforate anus do very well; those who have high type imperforate anus tend to have difficulty with fetal continence, which can vary in severity. This especially is true of infants who have absent sacral segments, so this problem should be identified early for the family. Other limitations of children who have imperforate anus depend greatly on the coassociation with other VATER anomalies.

**SUGGESTED READING**


**PIR QUIZ**

7. A 4-year-old girl presents with a 3-week history of a slowly enlarging lymph node in her neck. On examination you find a 4-cm tender lymph node in the left anterior cervical region. Although the overlying skin is reddened, the girl is afebrile and does not appear ill. Findings on the remainder of the physical examination are unremarkable. Aside from cat scratch disease, you should consider in the differential diagnosis each of the following conditions except:
   A. Atypical mycobacterial infection.
   B. Bacterial adenitis.
   C. Infected branchial cleft cyst.
   D. Kawasaki disease.
   E. Tuberculous adenitis.

8. Cat scratch disease is transmitted in each of the following ways except:
   A. Cat bite or scratch.
   B. Dog bite.
   C. Lick by a cat of injured skin.
   D. Person-to-person.

9. Manifestations of cat scratch disease may include each of the following except:
   A. Chronic tender regional lymphadenopathy.
   B. Diffuse dermatitis in patients who have human immunodeficiency virus infection.
   C. Encephalopathy.
   D. Erythema chronicum migrans.
   E. Parinaud oculoglandular syndrome.

10. Although use of cat scratch antigen for skin testing remains controversial, each of the following may help establish a diagnosis except:
    A. An elevated erythrocyte sedimentation rate.
    B. Characteristic lymph node biopsy.
    C. History of contact with a cat or a kitten that resulted in dermal or eye injury.
    D. Negative bacterial culture of pus aspirated from a suppurated lymph node.
    E. Regional lymphadenopathy developing 2 weeks after a cat scratch or bite.

11. Each of the following statements about the treatment of cat scratch disease is true except:
    A. Antibiotic therapy is generally not required in cases of cat scratch disease.
    B. For the majority of children, no therapy is required.
    C. Incision and drainage may accelerate recovery in selected cases.
    D. Needle aspiration of suppurated nodes may relieve pain.
    E. Surgical excision may be required if the diagnosis is in doubt.
Support Groups
Gay and Lesbian Parents' Coalition International (GLPCI)
PO Box 50360
Washington, DC 20004
(202) 583-8029

National Coalition of Gay Fathers
146 East 30th Street
New York, NY 10016

Children of Lesbians and Gays Everywhere (COLAGE)
2023 North Clark, Box 121
Chicago, IL 60657

Note: This list has been limited to national support groups that can assist physicians and families in locating local support and information services in their own or neighboring communities.

SUGGESTED READING
Professional Articles

General References on Gay and Lesbian Issues
For Adults and Adolescents
Alpert H. We Are Everywhere: Writings By and About Lesbian Parents. Freedom, Calif: Crossing Press; 1988
Rafkin L. Different Mothers: Sons and Daughters of Lesbians Talk About Their Lives. Pittsburgh, Penn: Cleis Press; 1990
For Young Children

PIR QUIZ
12. The data about children whose parents are gay or lesbian indicate that they are more likely to:
A. Be sexually abused.
B. Be socially isolated.
C. Become homosexual themselves.
D. Develop human immunodeficiency virus infection.
E. Have emotional problems.
13. In comparison with heterosexual parents, gay or lesbian parents have been shown to be all of the following except:
A. Less authoritarian.
B. More accepting of diversity.
C. More consistently nurturing.
D. Unlikely to maintain consistent contact after separation.
14. Examples of ways in which pediatricians can help support families in which the parents are gay or lesbian include all of the following except:
A. Assist in clarification of legal responsibilities.
B. Insist that parents disclose their sexual orientation.
C. Offer to talk with child care and school personnel.
D. Provide informational pamphlets and reading materials in the waiting room.
E. Review office registration and history forms for evidence of heterosexism.
vides most of the protein intake, the patient must receive enough phenylalanine for growth and development from measured amounts of protein-containing natural food, and blood phenylalanine must be monitored to ensure that it is neither too high nor too low. Special formulas never should be started before the diagnosis is confirmed and without supervision by a nutritionist who has training and experience in inborn errors. Only in this way can under- and overrestriction of essential nutrients be prevented. Compliance is a life-long issue for the patient and family, and best results are obtained when the primary care physician is in close contact with a group that can provide nutritional support.

Commercial formulas also are available to treat maple syrup urine disease, methylmalonic acidemia, and other disorders, but management of these patients is even more difficult. Intercurrent illnesses that cause metabolic decompensation must be prevented when possible (eg, early flu vaccines) and catabolism avoided by admission to hospital for intravenous glucose at the first sign of decreased intake.

Genetic counseling is another important aspect of management, and discussion of recurrence risk is essential. The number of disorders for which prenatal diagnosis (and carrier detection) is available is increasing constantly, as is the number of conditions in which knowledge of the molecular defect affects the manner in which testing is done and the accuracy of the results. As with treatment, it is increasingly difficult to impart the necessary counseling information without input from a unit that specializes in these conditions.

Conclusions
Diagnosis in the newborn who is acutely ill with an inborn error of metabolism is not very difficult and requires access to only a few laboratory procedures. Although some of these, such as organic acid analysis, are available in only a few specialized laboratories, the existence of reliable commercial air transport services has made such analyses available even to patients in remote areas. Increased awareness of these conditions and better use of laboratory resources should permit more affected newborns to be diagnosed and treated and proper genetic counseling to reduce morbidity and mortality.

SUGGESTED READING
Nyhan WL. Abnormalities in Amino Acid Metabolism in Clinical Medicine. Norwalk, Conn: Appleton-Century-Crofts; 1984

PIR QUIZ
15. You have just received the results of the screening program for congenital hypothyroidism for one of your newborn patients. The T4 concentration is low and the TSH level is elevated. These findings suggest:
A. Laboratory error.
B. Prematurity.
C. Primary hypothyroidism.
D. Secondary hypothyroidism.
E. Thyroxine-binding globulin deficiency.

16. You are asked to see a 5-day-old boy who has become increasingly lethargic and unresponsive. He is severely acidotic and has hyperammonemia. You suspect a metabolic disorder. Each of the following would be appropriate management except:
A. Dialysis.
B. Intraocular administration of arginine.
C. Intraocular administration of bicarbonate.
D. Intravenous administration of glucose.
E. Intravenous administration of vitamin B12.

17. Liver transplantation is the only approved effective treatment for which of the following:
A. Carnitine deficiency.
B. Glycine encephalopathy.
C. Hereditary tyrosinemia.
D. Isovaleric acidemia.
E. Maple syrup urine disease.

18. The mode of inheritance for most metabolic disorders in the newborn is:
A. Autosomal dominant.
B. Autosomal recessive.
C. Multifactorial.
D. X-linked dominant.
E. X-linked recessive.
increased psychopathology in the child—a vicious cycle. Parents must be guided and helped in these potentially disastrous situations.

Parents also should be guided to prevent making their children overprotected and overdependent. The child who has epilepsy should be provided a clear understanding of the nature of his or her problems.

All of these factors should be considered when making choices concerning the administration of anticonvulsant drugs (such as which ones to use, whether they should be used at all, whether dose changes should be made, and whether anticonvulsants should be stopped) and the general management of children who have epilepsy.

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Comment: Children who have a chronic condition often are subject to overprotection from family and some “labeling” as a vulnerable child. The impact of this family dynamic may impair overall optimum development and will need to be modified carefully. Children who have a chronic illness and are monitored by the pediatrician should attend carefully to these more subtle consequences of having a diagnosis of epilepsy (or any chronic condition for that matter) in addition to the more frequently discussed medical complications. Often this will mean additional discussion with the child and the parent or parents together. One of the most important targets of anticipatory guidance is children who have a chronic illness and their families.

Steven P. Shelov, MD
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