of 14 with symptoms similar to those which Patient A. S. developed at 11½ years of age; the youngest child, at the age of 8, had minimal electrocardiographic findings.

Incomplete development of the external genitals and mental retardation have been reported with several entities, some of which have an associated congenital heart defect. Our three cases differ from these entities because of the conspicuous lack of associated phenotypic or chromosomal anomalies, and the presumptive evidence of a cardiomyopathy of late onset. Cases of familial cardiomyopathy have been reported, but there has been no associated mental retardation or anomalies of the external genitals.

The occurrence of these anomalies in three siblings suggests a genetic etiology. The consanguinity of the parents favors an autosomal recessive pattern of inheritance.

REFERENCES

Intravenous alimentation in patients with cancer and neutropenia

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The successful delivery of hypertonic glucose-amino acid solutions intravenously provides effective nutritional support for patients when gastrointestinal alimentation is impossible or impractical. Obviously, the clinical application of such a procedure encompasses a variety of situations. In the management of children with acute leukemia and solid tumors, we encountered a number in whom provision of adequate nutrients and gastrointestinal bypass would have been advantageous.

In view of the reports of infectious complications of intravenous alimentation, the use of this technique in children with cancer, neutropenia, and impaired resistance to infection was undertaken with considerable caution. However, relatively few problems were encountered, and the usefulness of this technique prompted us to relate this experience.

METHOD
All patients with a primary neoplastic disease or neutropenia and who received intravenous alimentation between October 6, 1970, and November 6, 1971, were reviewed. Central venous alimentation was instituted only after severe irreversible weight loss not responding to dietary management. It was not used in terminal illness and considered only when the nutritional condition was life threatening or prevented the benefit from other therapy (surgery, full-dose chemotherapy, radiation). Intravenous alimentation was defined as delivery of a hypertonic glucose-amino acid solution intravenously by means of a central venous catheter. In all instances but one (Patient S. S., first course) percutaneous subclavian vein catheterization was used. No single intravenous catheter was left in place for longer than 30 days. A 0.22μ pore size Millipore filter was always used as a terminal sterilizing mechanism in the in
fusion line. Solutions were prepared under aseptic conditions in a laminar flow hood and provided 100 to 125 per cent of the caloric needs. Amino acids contributed at least 10 per cent of the calories. Sodium, potassium, chloride, phosphate, magnesium, calcium, and vitamins were added as commonly recommended, and their concentrations adjusted if indicated by their respective serum concentrations. Electrolytes and blood pH were regularly monitored: twice daily initially and every two days later. Particular attention was given to any possibility of infectious complications, i.e., fever, microbial isolates from blood or catheter tip cultures, or clinical suspicion of infection. Patients were monitored frequently with cultures for bacteria and fungi.

**RESULTS**

Nine patients received intravenous alimentation during the specified time. Details regarding each patient are shown in Table I. There were four males and five females. The patients with lymphosarcoma had primary lesions of the anterior mediastinum with involvement of the central nervous system. All except the patient with congenital neutropenia had been or were currently undergoing antineoplastic chemotherapy. In only two patients (S. S. and C. L.) was the primary disease under some degree of control (both were leukemia patients in hematologic remission and without other evidence of active leukemia).

Total patient days of intravenous alimentation was 168. One patient received only two days of therapy, whereas the patient with congenital neutropenia had four courses for a total of 67 days.

Complications of the procedure were encountered in two patients. Patient S. S. had persistent fever despite antibiotic therapy; the central venous catheter was removed and defervescence immediately followed. Subsequently *Staphylococcus aureus* was cultured from the catheter tip. Blood cultures were sterile. Patient D. L. had a febrile course just prior to his death; cultures revealed *Candida parapsilosis* in the blood and *Candida tropicalis* in the urine. Autopsy cultures from the lung, spleen, blood, and central venous catheter grew *Candida tropicalis*, and multiple organ invasion by Candida organisms was demonstrated histologically.

No significant problems with fluid and acid-base balance were encountered.

In five cases central venous catheters were not cultured; however, in these patients no infection was clinically apparent or suspected and cultures of the blood were sterile for bacteria and fungi.

Body weight either remained stable or increased slightly during parenteral alimentation.

**DISCUSSION**

The patients presented represent a group of individuals highly susceptible to nosocomial infections. Patients with diseases asso-
associated with granulocytopenia or impaired immune response characterize this group. More specifically, an increased incidence of opportunistic fungal infections was noted in patient populations comprised largely of individuals with neoplastic disease. Moreover, Curry and Quie\textsuperscript{2} reported a significant incidence (16 per cent) of fungal septicemia in a prospective survey of patients receiving parenteral alimentation.

It is therefore of note that in our group of patients receiving a total of 168 days of parenteral alimentation, there were only two instances of infectious complications and that one of these subsided readily upon removal of the infected catheter. Preparation of solutions in a laminar flow hood, the use of terminal Millipore filtration, and meticulous supervision of the procedures by the same physicians are important to the success of intravenous alimentation. The percutaneous method for subclavian vein catheterization may be less vulnerable to acquisition of infection than other methods.\textsuperscript{6}

In any group of patients with deficits of the defense mechanism, either inherent or as a result of neoplasia and chemotherapy, instances will arise in which malnutrition aggravates significantly the susceptibility to infection, and nutritional restoration will be lifesaving. Yet, the physician may be reluctant to use central vein alimentation for fear of infectious complications.\textsuperscript{5}

The conclusion from our experience is that parenteral hyperalimentation in selected cancer patients can be carried out with reasonable safety from infection provided meticulous attention is given to composition and delivery of the fluid. In all cases the decision whether or not to utilize intravenous alimentation must include weighing the possibility of complication against the potential benefit to be gained.

REFERENCES