Clinical Practice and Therapeutics

VACTERL/VATER association—Can a patient with VACTERL association live independently?

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A newborn baby boy with a maternal history of polyhydramnios noted during antepartum examination was found to have drooling, respiratory distress, and multiple congenital malformations soon after birth. An oral–gastric (OG) tube could not be inserted. Physical examination showed no dysmorphic facial features, a regular heartbeat without murmur, normal upper limbs, normal spinal curvature, penile hypospadias, and imperforate anus (Fig. 1A). Chest radiography revealed normal vertebrae with extra ribs, coiling of the OG tube and a distended gastric bubble, with a Gross classification type C tracheoesophageal fistula (Fig. 2). Whole body sonography demonstrated corpus callosum dysgenesis, a mild patent foramen ovale, and no renal anomalies. End-to-side anastomosis of the esophagus, repair of the trachea, and an anorectoplasty (Fig. 1B) were done at 4 hours postpartum. The patient was stable postoperatively. Feeding with breast milk was started through the OG tube on postoperative Day 7. The anal sutures were removed 13 days postoperatively, and anal dilatation was done daily. The patient was discharged 14 days postoperatively after the parents learned care techniques. At the time of writing, the patient was aged 5 months. After regular rehabilitation, his growth and development were all within normal limits, and his rehabilitation course will be finished shortly.

For primary care medical personnel, there are several major concerns in a child born with a congenital anomaly. These include its severity and whether it is life-threatening, isolated or combined with other anomalies, part of a pattern recognized as a syndrome, or associated with other anomalies. Clinicians need to determine whether any molecular genetic testing is currently available. Care by an interprofessional medical team and the parents’ psychosocial concerns and emotional response must also be considered. Explanation of outcomes and genetic counseling to evaluate recurrence risks are mandatory.

For patients, early infant stimulation programs, regular follow-up of neurocognitive milestones, a complete vaccination program, membership in a disease association, and lifelong medical care plans are needed.

The first concern of most parents with a child with a congenital anomaly is how well that child will function in life; for those with
VACTERL, the prognosis can be relatively positive. Patients with VACTERL association can live independently.

**Further reading**