Taking into account the experience of past workers(3) we omitted radiotherapy which is usually a part of the central nervous system directed therapy in our protocol (*Fig. 4*), though we continued to give him intrathecal methotrexate with a 25% reduction in dosage. Neurologically patient remained stable during remission. Despite a 25% reduction in the chemotherapeutic dosage our patient did achieve complete remission and remained in remission for 5 months. However, he developed recurrence of the mediastinal mass and succumbed to progressive disease after six months of diagnosis.

More information is required regarding dosage of chemotherapeutic agents so as to prolong remission in these patients.

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Acephalus Acardia Syndrome

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Acephalus acardia syndrome is characterized by absence of head and heart with abnormality of limbs. The condition is extremely rare and is estimated to occur once in 40,000 deliveries(1). It is exclusively seen in monozygotic twin pregnancies where one fetus develops normally while the other becomes an amorphous mass with its blood supply coming from the normal fetus through anastomosis in

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umbilical vessels in a monoamniotic placenta. We report a twin pregnancy in which one fetus had acephalus and acardia.

Case Report

A 35-year-old third gravida was admitted with labor pains in maternity ward and delivered a healthy female baby weighing 2.75 kg with Apgar score of 8 at one minute. After delivery of the first baby, she delivered spontaneously a second fresh still born female baby weighing 1.05 kgs. with multiple congenital malformations (*Fig.*). The head was constituted by a 3 × 3 cm soft

Fig. Showing absence of development of head, neck, upper limbs, left lower limb and right foot.

cystic bud with no demarcation of eye, ear, nose or mouth; the neck was represented by four small nodules. Thoracic wall was not properly developed; the ribs, sternum and scapulae were not felt. The trunk measured 22 cm in length. The umbilical cord was attached at junction of upper and lower trunk. Both the upper limbs were absent and were represented by 2 mm buds on either side. The female external genitalia and anus were well developed and were on left side.

The right thigh and leg were well developed but right foot and left lower limbs were absent.

Infantogram showed absence of development of the skull, upper limbs, scapulae, left lower limb and the right foot. Marked scoliosis of the spine was seen. The lungs were not aerated.

The placenta was monoamniotic monochorionic weighing 800 g. There was anastomosis between the umbilical arteries of the two lobes.

Discussion

The acardiac anomalies have been principally categorized according to the gross appearance. Based on the most widely adopted classification of Das(2), the acardiacs are divided into 4 groups:

1. *Acardius acephalus* is the most common type. They are headless and normally lack the thoracic organs. The intraabdominal organs are rudimentary. The upper extremities may or may not be present. The present case fits in this category.

2. *Acardius anceps:* This is the most developed form, characterized by partially developed head and face. The body and extremities are present except the heart. Ko *et al.*(3) have reported three cases of acardius anceps recently.

3. *Acardius amorphous:* There is only shapeless mass containing no recognizable organs.

4. *Acardius acormius:* This is the rarest form with the development of head only.

Although radiologic study or pathologic examination is necessary for a reliable classification(4-5), prenatal sonography is a useful and practical aid in classification(3). The etiology of acardiac anomaly remains unknown. The "twin reversed arterial perfusion (TRAP)" theory proposed by Van Allan seems the most widely accepted to date(6).

The acardiac twin, thus can be designated as "perfused twin", and the cotwin as "pump twin", based on the direction of the umbilical blood flow. Such reversal of blood flow depends on a relatively more powerful functioning heart in the normal cotwin(6). The acardiac anomalias, with very rare exception affect more severely the upper limbs or entire acephalic pole than the lower limbs(6). This is believed to be related to the retrograde blood flow along the umbilical artery to the abdominal aorta via the iliac artery. Therefore, the lower part of the body preferentially obtains more blood, although deoxygenated and nutrients-deprived, than the upper part(6). Normal cardiac formation depends prima-rily on the magnitude and direction of the mechanical hemodynamic forces.

In experimental animals, proper folding of the cardiac tube development depends on normal blood flow and pressure. Consequently, such reversal of blood stream results in disruption on early cardiac development or causes degenerative change later(6,7). Potter(8) suggested that the acardiac fetus represents primary agenesis of the heart, while others thought it to be a part of a generalized chromosomal defect.

As the "pump twin" can develop congestive heart failure, early antenatal detection of the condition is important. Harger *et al.*(9) found elevated amniotic fluid alpha-feto-protein levels in a holoacardium amorphous (other name of acardius amorphous) twin. Platt *et al.*(1) diagnosed two cases of acephales acardius by ultrasound and recommended surgical procedure of clamping cord of acardius fetus to prevent heart failure in the normal twin. Although few cases of acardia with multiple pregnancy have been reported from India(10-12), only one case of acardiac acephalic monster has so far been reported(13), from India.

Hence, this condition must be kept in mind by all obstetricians where in a multiple pregnancy heart of one twin is not detectable in ultrasound.

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Blood Transfusion in Autoimmune Hemolytic Anemia— A Practical Problem

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Autoimmune hemolytic anemia (AIHA) is characterised by pallor, icterus, hepatosplenomegaly and a positive Coombs' test. The overall incidence of the condition is 1 : 80,000(1), but the incidence in children is much lower since the majority of patients reported were over 40 years of age(2). We present a case of AIHA and highlight the problems faced while transfusing blood in this patient.

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Case Report

An 11-year-old girl was admitted with progressive pallor for 3 months and breathlessness for 15 days. There was no history of fever, anorexia, drug intake or injections, recent blood transfusion, joint pains or urinary complaints. Examination revealed pallor, icterus and signs of congestive heart failure with hepatomegaly of 5 cm and splenomegaly of 3 cm. There was no lymphadenopathy or petechial spots.

Investigations revealed a hemoglobin level of 3 g/dl, reticulocyte count of 45%, normal total and differential leucocyte and platelet counts. Peripheral smear was suggestive of severe hemolytic anemia, direct Coombs' test was positive, hemoglobin electrophoresis was normal and tests for G-6PD deficiency were negative. The serum bilirubin level was 3 mg/dl predominantly conjugated with normal levels of liver enzymes, immunoglobulins and complement. Antinuclear factor was negative and chest *X*-ray showed cardiomegaly. A diagnosis of idiopathic AIHA was made. The patient received three blood transfusions of 'O' Rh positive blood and prednisolone in the dosage of 2 mg/kg/day. Follow-up revealed rising hemoglobin levels and steroids were tapered to 5 mg/day when hemoglobin reached 10 g/dl.

Four years later, the child was again admitted with pallor and breathlessness. Examination showed pallor and features of congestive cardiac failure. She had lost her previous records and stopped steroids for