

Successful Management of Pregnancy in Arthrogryposis Multiplex Congenita

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Citation

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Abstract

Background:

Arthrogryposis multiplex congenita (AMC) describes the presence at birth of fixed joint contractures. This case of AMC in pregnancy highlights the potential problems encountered when managing the obstetric care of these patients.

Case:

A 27-year-old primigravida with known AMC since birth presented at 9 weeks of amenorrhoea. Care plan in pregnancy included monitoring of fetal growth by serial ultrasound scans from 24 weeks and maternal assessment using regular pulmonary function tests. Deterioration in maternal cardio-respiratory function dictated an elective preterm delivery under general anaesthesia balancing the maternal and fetal needs.

Conclusion:

A multidisciplinary approach involving anaesthetic, neonatal, obstetric and midwifery staff, in the antenatal and postnatal care of women with AMC is vital. Timing of delivery can be challenging in view of maternal risks especially when the fetus is healthy. It also emphasises the importance of forward planning as a key to a successful outcome of pregnancy in this condition.

CASE REPORT

A 27-year-old Asian woman attended the antenatal clinic at 9 weeks amenorrhoea. She herself had been delivered normally at term with AMC being diagnosed at birth. AMC affected the function of all her limbs, rendering her a moderate degree of disability. She had numerous orthopaedic operations on her limbs under general anaesthetic till the age of 14 years with no significant problems. She was also diagnosed as child with asthma and took regular doses of inhaled steroids and beta agonists. She had a paternal female cousin who suffered from same condition. There was no consanguinity.

On examination, her BMI was 31, she was 3ft (0.91m) tall. Spinal involvement produced a marked lumbar lordosis and thoracic kyphosis with normal neck architecture and extension. Abnormally shaped scapulae impaired movement in her arms. She had bilateral callipers leading to little mobility in her legs. Examination of her cardiovascular system was normal. She was reviewed by regional genetics team, she declined prenatal invasive testing.

Due to maternal habitus leading to difficulty in clinical obstetric examinations, serial scans were performed from 24 weeks of gestation, the anomaly scan being normal. Possibility of deterioration in respiratory function leading to elective preterm delivery was discussed. She had an anaesthetic review in the second trimester where possibility of regional techniques was ruled out due to abnormal vertebral anatomy.

The woman was admitted at 30 weeks of pregnancy with worsening breathlessness at rest. Investigations revealed abnormal peak expiratory flow rates and evidence of infection on chest X-ray. Examination revealed the uterine fundus to be at the xiphisternum due to her short stature and spinal deformity. Antenatal corticosteroids were administered and dosage of inhaled agonist and steroid was increased along with chest physiotherapy. Elective caesarean section was performed under general anaesthesia without complication in view of deteriorating maternal condition at 31 weeks. She delivered a healthy, 1100gms baby boy, which needed stay in neonatal intensive care due to gestational age. She had uneventful post-operative recovery.

DISCUSSION

AMC, first described in 1905, is a rare congenital syndrome of neurogenic and myopathic abnormalities, affecting approximately 1 in 3000 births. The etiology is unclear and multifactorial such as mechanical, vascular and neuronal.¹ It is marked by a limited range of motion in one or more major joints in the human body. While there is no cure, treatment options include splinting of affected joints, physical therapy to improve flexion and range of motion, and surgery to help reposition severely affected joints and limbs.¹ Those not severely afflicted typically live relatively normal lives, adapting to specific situations as required by the patient's particular symptoms. Women with AMC are of normal intelligence and able to lead a good standard of living, although may experience some mobility problems. Therefore, expectations of motherhood should not be surprising. Little is published about the care of women with AMC in pregnancy, which provides specific challenges to the attending obstetrician.

Respiratory function is affected due to advancing gestation in normal pregnant women. This is more pronounced in women with AMC due to lack of available space in the abdomen for uterine enlargement as well as diminished pulmonary reserve contributed by spinal deformity. This was the case in our patient where preterm delivery had to be planned in view of deteriorating maternal respiratory condition. We suggest monitoring of cardio-respiratory function from 28 weeks of gestation utilising pulmonary function tests and echocardiography in asymptomatic pregnant women with AMC. Serial ultrasound scans from 24 weeks were performed to monitor the fetal growth.

The risk of thromboembolism in these women is theoretically greater than in normal pregnancy due to the added risk of poor mobility due to joint affection. Prophylaxis against venous thromboembolism should be considered in women with AMC and should be continued after operative delivery.² Prophylaxis was not prescribed in this case as the patient's mobility was exceptional for her condition. Another potential difficulty was the patient's diagnosis of the reversible airways disease, asthma. The timing of delivery can be challenging especially when tests for fetal wellbeing are normal and the need for delivery is due to a serious deterioration in the maternal condition.

Anaesthesia-related difficulties present special difficulty in these patients, and early referral to anaesthetic colleagues is essential. Lower spinal deformalities, such as lumbar lordosis in this case, make regional anaesthesia a risky procedure. The additional risks of general anaesthesia for operative delivery must be weighed against the chance of successful regional anaesthesia. Abnormal vertebral anatomy can complicate the siting and action of epidural and spinal blocks as was the case in our patient. Women with AMC will usually undergo numerous operations under general anaesthetic in childhood, and review of the anaesthetic notes will be useful. Significant deformities of the upper spine and neck can make standard endotracheal intubation impossible and alternatives, such as fibre optic intubation must be explored.³ This is best achieved with elective operative delivery with input from neonatologists in the event of a preterm neonate.

The social circumstances of the woman can be challenging in their management and require a comprehensive approach to planning for care of mother and baby during and after delivery. In the above mentioned case, special arrangements with regards to caring for the baby after delivery were made. Social service provision, in terms of carers and housing is often needed. The planned caesarean section at 31 weeks allowed her to make the necessary arrangements in advance and liaison between obstetric staff, community midwifery and social services, which allowed a discharge plan to be put in place.

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