CLINICAL ARTICLE

Reconstruction of Limb Deformities in Patients with Thrombocytopenia-absent Radius Syndrome

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Objective: Developmental abnormalities of the appendicular skeleton are among the most common and easily identified birth defects. The aim of this report was to describe the phenotypic characterization of several patients with thrombocytopenia-absent radius (TAR) syndrome and the orthopaedic interventions performed on them. TAR syndrome is inherited in an autosomal recessive manner and results from compound heterozygosity of *RBM8A* mutations.

Methods: Reconstructions were designed and performed in five patients with TAR syndrome, mainly comprising orthopaedic interventions to correct their upper limb defects. Additional lower limb deformities (severe internal rotation of the tibiae) was been encountered in one patient.

Results: The affected patients' wrists were re-aligned and stabilized and the musculotendinous forces around the wrist rebalanced to reverse the ulnar forearm bow.

Conclusion: Patients with TAR syndrome who receive optimal treatment can expect to return to most activities of daily living with some limitation of wrist extension and ulnar deviation and, of course, with a reduced total active range of digital motion.

Key words: Longitudinal radial deficiencies; Reconstruction; Thrombocytopenia-absent radius syndrome

Introduction

Thrombocytopenia-absent radius (TAR) syndrome is a relatively uncommon condition characterized by absent radii with the presence of thumbs and congenital or early onset thrombocytopenia that tends to resolve in childhood. These patients have congenital deformities of the forearm and hand (often bilateral) with the hand at right angles to the forearms, the thumbs are always present and there is hypoplasia of the muscles and soft tissues in the arms and shoulders. They have a hemorrhagic tendency caused by thrombocytopenia that is usually present at birth or shortly thereafter; episodes of thrombocytopenia tend to occur less frequently with

increasing age in most patients^{1,2}. Radiologically, the radii are absent bilaterally with or without other upper limb abnormalities, the ulnae are short and malformed, the humerus is absent in 5%–10% of cases in whom the digits arise from the shoulder. In some patients, the digits are hypoplastic and the phalanges fused or both. Hypoplastic or absent middle phalanx of the fifth digit is a not uncommon feature. Shoulder abnormalities are diverse, including absent or hypoplastic glenoid fossa, acromion, scapula, or clavicle or a combination of these. Lower limb abnormalities include hip dislocation, phocomelia, coxa valga, genu varum, subluxated knee, hypoplastic or absent patella and patellar dislocation³. Additional deformities in

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Fig. 1 Clinical phenotypes of three children aged (A) 8 months, (B) 2 years and (C) 5 years. See text for details.

connection with thrombocytopenia have been reported, including femoral and tibial torsion, abnormal tibiofibular joint, club foot deformity and abnormal toe placement, Mullerian agenesis, horseshoe kidney, oesophageal atresia and others⁴⁻⁷. Hypoplasia or absence of the radius has been classified into four grades by Bayne and Klug⁸. Type I is characterized by late appearance of the distal radial epiphysis, type II by a small radius with proximal and distal epiphyses. In Type III there is a small proximal radius and in type IV complete radial aplasia.

Patients and Methods

The study protocol was approved by the Medical University of Vienna (Ethics Committee, EK Number 921/2010) and informed consent was obtained from the patient's guardians. This study involved clinical and radiographic evaluation of a group of children with TAR syndrome (three boys and two girls; age range 2 months to 4 years).

Congenital longitudinal radial deficiencies are frequently associated with other malformations. This is explained by the fact that many organs develop at the same time as the upper limb buds. It therefore behooves the orthopedic surgeon to be aware of these associations and to ascertain whether a patient has any serious anomalies that would make surgery hazardous and inadvisable. The initial evaluation of a child with radial longitudinal deficiency should include upper limb, spine, pelvis and lower limb radiography (skeletal survey to detect associated abnormalities). Doppler echocardiography, renal and pelvic ultrasound, and complete blood count are mandatory. Chromosomal tests are warranted, specifically to exclude Fanconi anemia. Most of our patients had serious functional disability because their abnormalities were bilateral, notably having difficulty performing activities of daily living such as dressing, feeding, and washing. Limitations in elbow flexion increased their functional impairment. Our patients' deficiencies were compatible with type IV of Bayne and Klug's classification of radial longitudinal deficiency. Their birth weights and length were around the 10th percentile. All patients had bilateral radial

club hands associated with various degrees of multiple petechial hemorrhages in the inguinal region, on the medial surface of the thigh and to a lesser degree over the abdomen, back and buttocks. They had markedly reduced platelet counts (thrombocytopenia) associated with melena in infancy (thrombocytopenia $<40,000$ platelets/mm³). The genotypic confirmation was obtained in three families who manifested the condition in an autosomal recessive manner; their deformities resulted from compound heterozygosity of *RBM8A* mutations.

Three of the children (aged 8 months [Fig. 1A], 2 years [Fig. 1B] and 5 years [Fig. 1C]) had clinical phenotypes characterized by brachycephaly, micrognathia, frontal bossing, large frontal area, depressed nasal bridge, faint eye brows, micrognathia and full cheeks (Figs. 1A,B). Their upper limb deformities comprised preservation of the thumbs together with bilateral absence of the ulnae and deformities associated with bilateral humeral dislocation. They also had hypoplasia of the muscles and soft tissues in the arms and shoulders (Figs. 1A–C). For logistical reasons, it was not possible to perform genetic tests in the remaining two patients; however, they manifested the full phenotypic features of the TAR syndrome.

Radiological Abnormalities

All studied patients had congenital complete absence of the radius with preservation of hypoplastic thumbs. They therefore lacked radial skeletal support of the carpus and had severe soft tissue contracture on the radial side of the forearm. The marked deviation of the hands was notable, they almost formed an angle of 90° with the forearm. When the arm was flexed, the upper arm lay directly against and parallel with the forearm with its radial border touching it. This overall radiographic phenotype was compatible with type IV radial deficiency (Fig. 2). Deletion/duplication analysis was performed to identify a 200 kb minimally deleted region in chromosome band 1q21.1. The presence of this deletion was sufficient to verify the diagnosis of TAR syndrome in these patients.

Fig. 2 Congenital absence of the radius in two different patients with preservation of hypoplastic thumbs, complete absence of the radius (Type IV). Please see text for details.

Surgical Procedures

All patients had absent radii (type IV), the radii being completely absent, the hands severely radially deviated and the forearm short. The hands were fixed in pronation and they substituted wrist flexion for supination. Corrective surgery can realign such limbs, but cannot reverse the anatomical and the biomechanical defects.

The following guidelines were followed with our patients.

1. Treatment was dependent on age.
2. Distracting the soft tissues of the wrist started in early infancy to maintain soft tissue length.
3. After the age of two months, parents were taught a stretching program that was combined with night splinting if possible. This aims to correct the position of the wrist and is normally planned for when the child starts to walk.
4. Between the ages of 6 months and 2 years, parents were asked to continue the stretching program, combined with night splinting if possible.
5. After the age of 3 years, splinting and stretching are not effective, the limbs having almost doubled in size since birth. This makes surgical intervention easier.
6. The surgical plan comprised the following procedures: i) centralization; and ii) radialization.
7. An Ilizarov correction was performed.
8. Maximal care was taken to protect the distal ulnar epiphyses and their blood supply.

Duration of Follow-up Time, Hand Function and Postoperative Complications

Follow-up is continued until adulthood when growth has been completed to confirm that the stability of the hand and arm are acceptable. Wrist motion will never be as full as in normal children; however, two of the children devised ways to perform everyday tasks and activities. In three children, radial deviation recurred because of deficient growth of the ulna and in

another child the ulna curved in connection with the missing radius (growth arrest and recurrence after early centralization). Therefore further surgeries are planned to correct these deformities.

Treatment

Because our group of patients had the most severe form of radius deficiency (type IV); our first approach was to stretch and splint the hands. This was followed by centralization of the carpus on the ulna, with tendon balancing or radialization of the ulna under the carpus. When deciding whether to centralize the carpus, a crucial factor is the range of motion of the patient's elbow. If the elbow joint is stiff in extension, any surgery that further restricts the patient's ability to reach the face with the hand is contraindicated. When a radiocarpal arthrodesis has been performed, compensatory motion occurs through rotation of the radio-ulnar joint. Because these patients cannot rotate their forearms, compensatory motion is restricted. Distraction lengthening of soft tissue may be useful for such radial deficiencies.

As a typical example of multistage management of bilateral upper limb deformities, we here present a case of 7-year-old girl with TAR syndrome to illustrate our surgical procedures.

The first step was fitting an Ilizarov frame on the left forearm and hand. An osteotomy of the ulna was performed with the goal of lengthening the left forearm. Lengthening of 4×0.25 mm a day was begun on the second postoperative day (Figs. 3A,B).

Four weeks later, the Ilizarov frame on the left side was removed and the carpus reradialized with additional fixation with two K-wires and a plaster cast.

Two months later, treatment began on the right side with revision and neurolysis of the median nerve and application of a mini-Ilizarov frame on the right hand; the K-wires were simultaneously removed from the left hand. Lengthening started on the second day postoperatively. Three weeks later, the mini-Ilizarov frame on the right side was removed and reradialization of the carpus performed by fixation with two K-wires and use of a postoperative cast for 6 weeks.

After another 4 months, an Ilizarov frame was fitted with inclusion of the hand and corticotomy of the proximal ulna for lengthening performed (start of lengthening on the fifth postoperative day with 3×0.25 mm distraction per day). The hand part of the Ilizarov frame was removed after 8 weeks and the rest of the Ilizarov frame and postoperative cast after 12 weeks.

At 6 months follow-up recurrence of the ulnar bowing on the right side was detected and an intramedullary nail implanted to restore the full length of the ulna. This procedure was accomplished by temporary arthrodesis of the wrist joint with K-wires and plaster cast. Six months later, full consolidation was detected and hardware removal recommended. Unpredictably however, removal of the intramedullary nail from the right ulna proved impossible because of massive bony fixation. Moreover, an intraoperative fracture of the proximal ulna occurred with the nail *in situ*. A cast was therefore fitted postoperatively.

Fig. 3 The first step of fitting an Ilizarov frame on the left forearm with inclusion of the hand has been performed, together with an osteotomy of the ulna was performed with the goal to lengthen the left forearm. Lengthening began on the second post-operative day at 4×0.25 mm a day.

Two and a half years later, an Ilizarov frame was fitted on the left ulna for lengthening. Lengthening started on the seventh postoperative day at 2×0.25 mm per day. The hand part of the Ilizarov frame was removed after 4 months and the rest of the Ilizarov frame 6 months after it had been fitted, together with fixation of the right ulna with a 10 cm reconstructive plate and plaster cast. A further 15 months later, transfer of the tendon of extensor digitorum was performed because of loss of motion in the left fingers and thumb secondary to external fixation with the Ilizarov frame (Fig. 4).

A further 6 months later, revision was performed: the right intramedullary ulnar nail with bone graft was removed and autologous bone material added to stabilize and re-lengthen the limb with the Ilizarov frame.

The Ilizarov frame was removed and reconstruction performed with a free fibular graft together with fixation with a reconstruction plate 5 months after the previously mentioned surgery (Figs. 5A,B).

Fig. 4 Results of multistage corrections (after the second lengthening with an Ilizarov frame of the ulnae).

In one patient with severe internal rotation of the right tibiae, a derotational tibial osteotomy was performed in the same procedure as acute external rotation. Fixation was achieved with a locking plate without postoperative casting (Fig. 6).

Discussion

The TAR syndrome is characterized by radial aplasia with preservation of the thumbs in combination with thrombocytopenia of early onset. Megakaryocytes are reduced and anemia, eosinophilia and leukemoid granulocytosis may be seen. If the infant survives, the hematological abnormalities become less severe with increasing age. Lower limb malformations, congenital heart defects, and abnormalities of the ribs and cervical spine may also occur¹⁻³.

The thumb is always present, as are hypoplasia of the muscles and soft tissues in the arms and shoulder.

Fig. 5 Radiographs showing the final correction in both forearms.

Fig. 6 Due to severe internal rotation of the right tibiae in one patient, a derotational tibial osteotomy was performed together with acute external rotation. Fixation was achieved with a locking plate without postoperative casting.

Hemorrhagic tendencies caused by thrombocytopenia are present from birth or shortly thereafter; in most cases the number and severity of thrombocytopenic episodes decrease with increasing age. The platelet counts are typically below $50 \times 10^9/L$ and can be low as $<10 \times 10^9/L$. The most common clinical presentations of the thrombocytopenia are gastrointestinal or intracerebral hemorrhage⁷.

Ballmaier *et al.* investigated the pathophysiology of thrombocytopenia focusing on thrombopoietin (TPO), the main regulator of thrombopoiesis, and its receptor⁹. Serum concentrations of TPO were high whereas expression of the thrombopoietin receptor (c-mpl) was similar to that of controls. An absence of *in vitro* reactivity to TPO was shown. The authors concluded that the defective platelet production is not the result of lack of TPO production, but of a lack of downstream response in the c-mpl signal transduction pathway. Strippoli *et al.* failed to find mutations in the thrombopoietin receptor gene (c-mpl)¹⁰. Letestu *et al.* found fewer colony-forming unit-megakaryocytes in the bone marrow than in normal subjects and that a proportion of the megakaryocytes

were unable to fully differentiate, suggesting that the defect lies in the early stages of megakaryocyte differentiation¹¹.

Greenhalgh *et al.* reported a clinical study of 34 patients with TAR syndrome, 47% of whom had lower limb anomalies, 47% cow's milk intolerance, 23% renal anomalies and 15% cardiac anomalies¹². Abnormalities not previously described in association with TAR syndrome included facial capillary hemangiomas, intracranial vascular malformations, sensorineural hearing loss and scoliosis.

The TAR syndrome is inherited in an autosomal recessive manner and results from compound heterozygosity of *RBM8A* mutations. Affected individuals typically have one *RBM8A* hypomorphic mutation along with a null mutation, usually a minimally deleted 200 kb region in chromosome band 1q21.1. Approximately 75% of probands have inherited the 200 kb minimally deleted region from an unaffected parent; the deletion occurs de novo in about 25% of probands. Siblings of a proband are at increased risk of also having TAR syndrome; however, because the minimally deleted 200 kb region in chromosome band 1q21.1 occurs de novo in about 25% of probands, the risk appears to be less than 25%. Albers *et al.* reported that 51 of 55 subjects with TAR syndrome had a 200 kb deletion on 1q21 whereas two had a truncation or frameshift (null) mutation in the *RBM8A* gene on one allele¹³. Of these 53 cases, all had one of two low-frequency single nucleotide polymorphisms in regulatory regions of *RBM8A* on the other allele. The likelihood of this mode of inheritance occurring by chance is less than 5×10^{-228} .

Radial deficiency is a relatively common upper limb abnormality that is associated with numerous known genetic and chromosomal conditions as well as with concomitant skeletal, visceral, cardiac, hematologic, renal and metabolic disorders. TAR, VATER (Vertebrae, Anus, Trachea, Esophagus and Renal), Holt–Oram, and Fanconi syndromes present with radial dysplasia^{13,14}. Klopocki *et al.* reported 30 unrelated patients with TAR and found a microdeletion at 1q21.1 in all. The deletion had occurred de novo in 25% of cases¹⁵. Inheritance of the deletion occurred equally from mother or father and it is postulated that the TAR phenotype develops only in the presence of an additional as yet unknown modifier (mTAR).

Radial longitudinal deficiency results from a failure of formation of the radial side of the hand, forearm, and upper arm. The condition was originally classified according to the extent of absence of the radius, but has since been extended distally to include carpal deficiency and hypoplastic thumb as well as a more severe deficiency affecting the upper arm¹³. Radial deviation of the wrist is the most common clinical manifestation of radial longitudinal deficiency. It can be corrected by a centralization procedure in which the hand and wrist are aligned onto the distal ulna to improve their appearance and possible function. This procedure is not universally accepted because recurrence is common and it may not improve function^{16,17}.

Decisions about treatment for radial deficiencies must take into account the prognosis of any coexisting conditions.

The classification of radial deficiency is based on the amount of residual radius present. In type I, there is a short distal radius. Type II consists of a small radius with proximal and distal growth plates, this form of deformity is uncommon. In type III, there is a small proximal radius. Type IV, which is the most common form of radial deficiency, comprises complete absence of the radius, often accompanied by absence of the radial carpi and first and second metacarpals. The elbow may be unstable and stiff or both because of severe soft tissue contractures on the radial side of the wrist (this form of deformity increase with age). The operative treatment of radial dysplasia aims to align and stabilize the wrist, rebalance the musculotendinous forces around the wrist and reverse the ulnar forearm bow. The use of an external fixator for soft tissue distraction has recently been promoted for better balancing the hand on the ulna. Although the short-term results are promising, the long-term outcomes are unknown^{8,18,19}.

Goldfarb *et al.* has described the indications, technique, and early outcomes of using a circular frame to achieve pre-centralization distraction²⁰. Sabharwal *et al.* reported the use of a circular frame for pre-centralization distraction in radial longitudinal deficiency²¹. Taghinia *et al.* suggested the use of a unilateral frame for pre-centralization distraction in severe and neglected radial deficiency²².

With optimal treatment, the patient can expect to return to most activities of daily living with some limitation of wrist extension and ulnar deviation and, of course, with reduced digital active range of motion. The large number of recommended procedures reflects the fact that surgery on the radial club hand is difficult and results sometimes unsatisfactory. The high failure rate is attributable to the fact that only the bones are considered while the muscles are ignored²³.

Buck-Gramko developed the technique of radialization (because the head of the ulna is positioned under the radial carpal bones and not at its center). This provides mechanical advantages, because radial positioning of the ulnar head and

transposition of the radial wrist muscles makes the lever arm to the ulnar side much longer and more powerful. The resultant mobility is better, because no carpal bones are excised. A further improvement in outcomes has been achieved by pre-operative distraction of the soft tissues around the wrist. This markedly facilitates subsequent surgery, avoids excision of carpal bones, and takes any pressure that would otherwise damage the growth plate to the distal end of the ulna^{24,25}.

We have described here five patients with TAR syndrome. Interestingly, the full clinical and radiographic documentation, including genetic tests, were performed retrospectively. In other published articles, the surgical treatment has focused on radial aplasia in general and this was also the main focus of our discussion. We performed extensive phenotypic and genotypic characterization of our TAR patients. From early in life, these babies were encouraged by the use of toys to use their affected hands. Despite the sometimes overwhelming distress parents feel in the face of their offspring's deformities, they have an unavoidable responsibility to help their children. They can deal with the crisis through fostering positive attitudes and feelings toward their offspring. Generally speaking, our patients were able to pick things up by using both hands together to grip them to their bodies and were able to grip some items between their fingers. However, two of the children hid their affected limbs and refused to use them.

The aim of this paper was to present the known causes of multiple limb malformations complexes in patients with TAR syndrome and to illustrate our orthopaedic reconstruction procedures for improving limb function.

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