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Research Article

Two-stage treatment of extremity deformities associated with thrombocytopenia-absent radius syndrome

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ABSTRACT

Objective: The aim of this study was to evaluate the results of 2-stage treatment of upper and lower extremity deformities in patients with thrombocytopenia absent radius syndrome.

Methods: Four patients (3 female, 1 male) with a mean age of 1.8 years (range 1-4) were included in the study. The patients were followed up for an average of 5.5 years. All 4 patients had bilateral radial longitudinal deficiency, whereas only 2 patients had bilateral fixed knee contractures. A 2-stage surgical procedure was implemented. The surgical procedure performed for radial longitudinal deficiency consisted of distraction with an Ilizarov frame in the first stage, followed by centralization performed in the second stage. Knee contractures were first treated using an Ilizarov frame, followed by a hamstring tendon transfer in the second procedure. Radiological evaluation of the radial longitudinal deficiency was done by measuring hand-forearm angle, hand-forearm position, and ulnar bowing preoperatively and at postoperative follow-ups. Knee contracture was evaluated by measuring the angle preoperatively and at postoperative follow-ups.

Results: The mean hand-forearm angle values of patients at preoperative assessment, early postoperative period, and at the last follow-ups were 82.60, 5,80, and 11.10, respectively ($P < .001$). The hand-forearm position values were −14.25 mm, +11, and +7.1 mm, respectively ($P < .001$). The ulnar bowing values were 7.3°, 4.5°, and 2.9°, respectively ($P < .001$). Recurrence of the radial longitudinal deficiency deformity requiring surgery occurred in 1 patient. In the other 3 patients, some deformity recurred but did not require surgical intervention. In addition, 1 patient with knee flexion contracture had a recurrence of the contracture that did not require surgical intervention. There was no circulatory disorder or skin necrosis in the lower or upper extremities of the patients.

Conclusion: This study has shown us that two-stage treatment is a reliable method for lower and upper extremity deformities accompanying thrombocytopenia absent radius syndrome. However, recurrence is still a major problem.

Level of Evidence: Level IV, Therapeutic Study

Introduction

Thrombocytopenia-absent radius (TAR) syndrome is very rare (1/100 000). Its diagnosis is based on 3 cardinal signs: thrombocytopenia, bilateral total absence of the radius, and the presence of the thumb. Extremity deformities in patients including cardiac (atrial septal defect and tetralogy of Fallot) (15%), gastrointestinal (cow's milk allergy) (47%), and urinary system (kidney location anomalies) (23%) abnormalities may also accompany.[1] Although thrombocytopenia is the main cause of mortality in the first months of life, it increases over time and even returns to normal in adulthood. Other systemic anomalies do not generally cause mortality, and the life span of the patients is similar to that of the general population.[1,2]

Limb anomalies can be seen in the upper and lower extremities. Radial longitudinal deficiency (RLD) due to the absence of the radius is the leading cause of upper extremity anomalies. The ulna is always deformed. The thumb may be present but dysplastic. Extensor muscles are atrophic or dysfunctional. The forearm muscles, which should attach to the radius, attach to the carpal bones and play an important role in radial deviation. Deformations ranging from aplasia to hypoplasia may occur in the humerus, glenoid, and scapular bones.[1]

Lower extremity abnormalities occur in 47%-80% of cases. Genu varum, coxa valga, tibial internal rotation, knee laxity, and flexion contracture are the most frequently reported deformities.[1,3]

Deformities in individuals with TAR syndrome should be treated both cosmetically and functionally because they complicate the patient's life. Our aim in the present study is to discuss the 2-stage treatment of RLD and knee deformities accompanying TAR syndrome with Ilizarov external fixators. We hypothesized that the 2-stage treatment of RLD and knee contracture accompanying the TAR syndrome will lead to lower rates of recurrence, less skin necrosis, less neurovascular injuries, and better correction of range of motion (ROM) in affected extremities.

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Figure 1. Correction of the radial longitudinal deficiency with an Ilizarov device.

Materials and Methods

Four patients diagnosed with TAR syndrome in our clinic with thrombocytopenia, bilateral absence of the radius, and presence of the thumb were included in the study retrospectively. Informed consent was obtained from the legal guardians of the patients before data collection. The laboratory, radiology, and examination data of the patients were obtained from the hospital automation system.

After the patients presented to our clinic, surgical intervention was scheduled considering the improvement of platelet counts, development of bones, and other systemic causes. Before the surgical intervention, consultation with the pediatrics and pediatric hematology departments was conducted. Surgical treatments of the patients who were followed up from birth were started to be planned when the patient turned 1 year old. A 4-year-old patient who had not undergone surgery before was operated on following the necessary preparations. Thrombocytopenia status was considered in terms of anesthesia.

When planning the treatment of both the upper and lower extremity deformities of the patients, 2-stage treatment was planned. At each stage, both the right and left extremities were operated on during the same session. In the first stage, gradual distraction of the soft tissues with an Ilizarov circular external fixator was followed by deformity correction and reconstruction in the second stage. Thus, attempts were made to prevent possible soft tissue necrosis problems and circulatory disorders.

HIGHLIGHTS

- Thrombocytopenia-absent radius (TAR) syndrome is a rare syndrome and the correction of the deformities are important aspects for the care of these patients. This study aimed to describe the 2-stage treatment of radial longitudinal deficiency and knee deformities accompanying TAR syndrome with Ilizarov external fixators.
- The results showed that all of the patients had improvements in the upper and lower extremity functions. No skin necrosis or circulatory problems was seen in the lower or upper extremities of the patients. However, pin-site infection was seen with both lower extremity and upper extremity Ilizarov frames in all patients although they were successfully treated with antibiotics and wound dressings.
- The results suggest that performing definitive operations following soft tissue distraction with external fixators is an effective method in the treatment of both lower and upper extremity deformities of TAR syndrome.

Surgical technique

The patients were evaluated by the pediatric and pediatric hematology departments before the operation, and their platelet counts were checked. Preoperative thrombocyte suspension was prepared for the patients. All surgical procedures except pin removal were performed under general anesthesia. A circular Ilizarov frame was placed in the patients in the first session without any soft tissue loosening. A half/full ring was placed on the metacarpal bones with stop wires. On the ulna, 2 full rings were installed using Schanz nails. Hand and ulnar rings were combined with appropriate hinge and motor units. From the third day after the operation, correction was started with 1 round per day. Then, after the fixator had remained in the upper extremity for a total of 6 weeks, the fixator was removed. Centralization with a dorsal incision was performed in the same session (Figure 1). Soft tissue release was performed on the radial side. It was fixed intramedullary to the ulna with a K wire by aligning the carpal bones and 2-3 metacarpal bones. The tip of the K wire was kept subcutaneously within the metacarpal bones. Follow-up was started with a long arm brace. The K wire was removed from under the skin with the help of local anesthesia 3 months later. In patients with knee contracture problems, an Ilizarov frame was established to contract the lower extremity in the same session. Two rings were installed on the tibia and 1 full and 1 half ring were installed on the femur. The rings were fixed to the tibia with stoppered K wires, a distal femur K wire, and a proximal Schanz nail. Hinges and a motor unit were placed. Contracture opening was started from the third day. It progressed with 2 tours a day. After correction was achieved in the sixth week, they were taken back to the hospital (Figure 2). In the same session as the fixator removal, the medial and lateral hamstring tendons were transferred to the quadriceps tendon. They were then followed with a long leg brace.

After RLD correction, the first 6 weeks were followed with a long arm brace (day and night). After the sixth week, a short arm brace was used during the day and a long arm brace was used at night. Movement of the fingers and metacarpophalangeal joint was allowed with the brace. After 3 months, they were allowed to make wrist movements. Mobilization was performed with a long leg walking device in patients who underwent knee contracture correction. The device was used day and night. After the 6-month checkup, the extremity was removed from the device and knee motion was started.

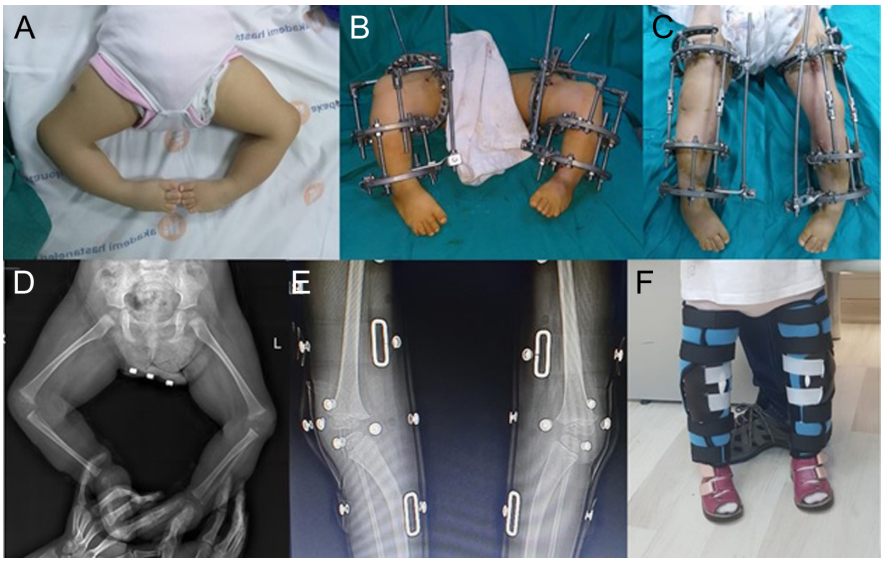

Figure 2. A-F. Gradual correction of knee-fixed flexion contracture with an Ilizarov device in the lower extremity. (A) Preoperative image. (B) Insertion of the Ilizarov contracture device. (C) Condition after the contracture was opened. (D) Preoperative direct radiograph. No hip dysplasia. There is no structural defect in the femur or tibia. Patella agenesis is present. (E) Standing x-ray taken after the correction was completed and the patient started walking without support with the brace. There was valgum in the bilateral tibia. (F) Standing clinical photograph of the patient with a brace.

Radial longitudinal deficiency objective grading of the patients was done using the radiological criteria hand–forearm angle (HFA), hand–forearm position (HFP), and ulnar bowing (UB) angle. In the radiological evaluation of the patients, HFA, HFP, and UB angles were measured on x-rays (Figure 3).[4] Early postoperative stage was defined as postoperative 3 months following removal of the external fixators and centralization. As control measurements, measurements performed at the last follow-up control were recorded. These measurement values were used both in preoperative and in postoperative checkups. In the clinical evaluation, measurements of ROM of wrists were performed by 1 surgeon. When evaluating lower extremity deformities, joint ROM, muscle strength, and general morphology were evaluated. The clinical evaluation was evaluated according to whether or not the patient was mobilized after surgery.

Statistical analysis

The changes in HFA (°), HFP (mm), and UB angle at the checkups were statistically compared before and after the operation. The obtained data were loaded into Microsoft Excel. Statistical Package for Social Sciences 17 was used in the analysis of the data (Chicago: SPSS Inc.). In the statistical evaluation, the median, mean, and SDs of measurement values were evaluated as ratios and percentages in the cross-sectional data. Evaluation in more than 1 dependent group was conducted with the nonparametric Friedman's test. The comparison of the values within the group was evaluated with the Wilcoxon rank test. The statistical significance level was set at $P < .05$.

Results

Four patients were included in the study. Three of the patients were girls and 1 was a boy. The mean age of the patients was 1.8.[1-4] Three of the patients were operated on at an average age of 12 months. The other patient was operated at the age of 4 years. The follow-up periods of the patients were 3, 4, 7, and 8 years, respectively (mean 5.5 years). The follow-up of 3 patients started after birth and of 1 patient at the age of 4 years. All patients were diagnosed by Pediatric Hematology units and referred to our clinic. The mean platelet values at the time of initial diagnosis of the patients were 53 750 (26 000-86 000) × 1000/mm^3, 122 000 (63 000-171 000) at the time of the operation and 216 500 (104 000-332 000) at the checkups (Table 1). One of our patients had leukocytosis (38 500).

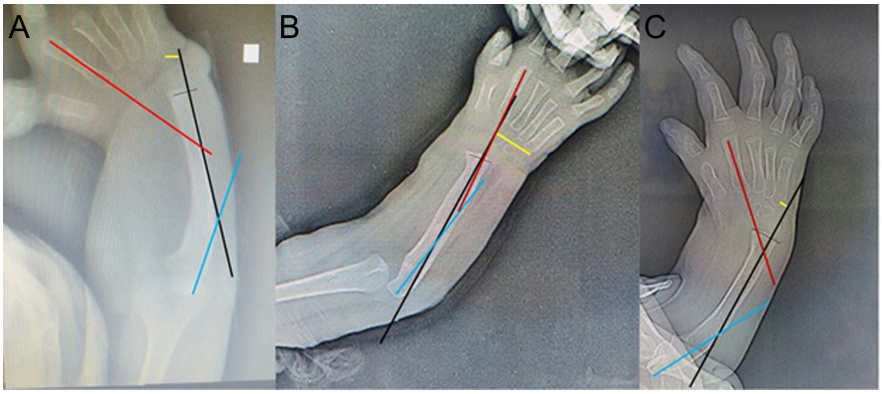

Figure 3. A-C. Radiological evaluation. Hand–forearm angle (red-black line), hand–forearm position (yellow line), ulnar bowing (blue-black line). (A) Preoperative, (B) early postoperative, and (C) checkup. There is some recurrence in the checkup.

Table 1. General demographics of patients

Patient number	Age	Gender	Control (years)	RLD	Lower extremity problem	Additional problem	Complication
1	1	Girl	8	Yes	None	Bilateral third to fourth intoe deformity	Bilateral recurrence of RLD and thumb reconstruction
2	4	Boy	7	Yes	Yes (bilateral contracture, internal tibial torsion, and absence of patella)	Bilateral short fourth metatars	Knee contracture progression and mild recurrence of RLD
3	1	Girl	4	Yes	None	Right thumb polydactyly Atrial septal defect	Mild recurrence of RLD
4	1	Girl	3	Yes	Yes (bilateral contracture, internal tibial torsion, and absence of patella)	Pelvic kidney	Mild recurrence of RLD

RLD, radial longitudinal deficiency.

Additional systemic problems were present at the time of diagnosis. Two patients had facial anomalies with wide foreheads and mandible hypoplasia. One patient had a pelvic kidney. One patient had an atrial septal defect. The defect closed spontaneously at 24 months of age.

All patients had bilateral absence of the radius, UB, and short forearms. Thumbs were present in 4 patients. One patient had unilateral thumb polydactyly. In this patient, polydactyly excision was performed during the centralization procedure in the second session. Elbow joints were mobile in all patients. There were no significant humerus, glenoid, or scapula deformations.

Two of our patients had flexion contractures in both knees, absence of the patella, and internal tibial torsion. These patients had quadriceps muscle contractions. One patient had bilateral short foot fourth metatarsal shortness and the other patient had bilateral third-fourth toe deformity (Table 1).

In the radiological evaluation, the mean preoperative HFA was 82.6°, the HFP was –14.3 mm, and the UB angle was 7.3°. At the last follow-up of the patients, these values were 11.1°, +7.1 mm, and 2.9°, respectively (Table 2). In the radiological evaluation, for early postoperative control, x-rays were taken in all patients at the third month. Control radiographs were taken at the last follow-up (8, 7, 4, and 3 years). Recurrence of the bilateral RLD deformity requiring surgical intervention occurred in 1 patient after 2 years. In this patient, recurrence started 6 months after the operation. Brace treatment was initiated at early period. However, surgical intervention was decided upon the progression of the deformity. This patient had instability in the wrist joint. Recentralization was applied to the wrist during the patient's revision surgery. The increase in HFA in the other 3 patients was considered mild recurrence, and surgical intervention was not considered necessary. Our patient who underwent revision surgery due to recurrence had the longest follow-up period. We continue to follow up, as other patients may relapse in the future.

Regarding clinical evaluations, wrist dorsal flexion was measured with a mean of 15.75 (6-23)° and volar flexion with an average of 24.25 (12-33)°. Average ROM of the wrist was 40 (18-51)°. Wrist movement limitation and extensor muscle insufficiency were present in all patients, but they were all able to use their hands. All patients and their families were satisfied with the cosmetic appearance of the hands at the last checkup.

The 4-year-old patient, who had not been able to walk before, started to mobilize with a brace in the third month after surgery. A 1.5-year-old patient started walking with a brace at 18 months and without support at 24 months after surgery. The 4-year-old patient had limited flexion of the knee. At the last checkup, the flexion contracture of the knee was 15° and 20°, but he could be mobilized. Although there was a preoperative contracture in patients whose bilateral knee deformity was corrected, we observed an instability problem after recovery. In order to solve this problem, the patients were mobilized with a brace.

Skin necrosis and circulatory disorders were not observed in the lower or upper extremities of the patients. Pin-site infection developed with both lower extremity and upper extremity Ilizarov frames in all patients. The infections regressed with wound dressing and antibiotics. No chronic infection developed in the pin base area after fixator removal.

Table 2. Preoperative, early, and late postoperative radiological and clinical evaluation of RLD

	Preoperative (1)	Early postoperative (2)	Control (3)	*P*
Hand–forearm angle (°)	82.625	5.750	11.125	.000335 * 1-2: .011** 1-3: .012** 2-3: .011**
Hand–forearm position (mm)	–14.25	+11.000	+7.125	.000335* 1-2: .012** 1-3: .012** 2-3: .011
Ulnar bowing angle (°)	7.250	4.500	2.875	.000912* 1-2: .027** 1-3: .011** 2-3: .027**
Wrist dorsal flexion	NA	NA	15.75′	NA
Wrist volar flexion	NA	NA	24.25′	NA

NA, not available; RLD, radial longitudinal deficiency.
*Friedman's test,**Wilcoxon rank test.

Discussion

The major problem faced by patients with TAR syndrome in the first year of their lives is thrombocytopenia. However, studies have shown that platelet counts increase in most patients over time and even return to normal limits.[1,2] In our patients, while the mean platelet value was 53 750 × 10^3/mm^3 at the time of diagnosis, it increased

Table 3. Preoperative, early postoperative, and control knee contracture angle values of the patients

Patient number	Preoperative knee	Postoperative knee	Control knee
1R	None	None	None
1L	None	None	None
2R	110	0	15
2L	110	0	20
3R	None	None	None
3L	None	None	None
4R	95	0	5
4L	95	0	10

to an average of 122 000 during the surgical procedure. In the most recent follow-up, the average was 216 500. In TAR syndrome, leukocytosis has been reported in addition to thrombocytopenia.[1,2] In 1 of our patients, the present picture was accompanied by leukocytosis (white blood cell: 38 500).

It is recommended that the timing of extremity surgery should be between 1 and 2 years of age in patients with TAR syndrome.[1] During this period, both the bone development of the patients progresses and the thrombocytopenia may regress. Thus, the external fixator is applied more easily and the need for platelet transfusion is reduced.

Two basic approaches have been described in the treatment of RLD: single-stage centralization with external fixation and post-distraction.[4-6] However, the disadvantages of single-stage surgery are circulation problems in the skin or extremities due to acute distraction, shortening of the ulna for reduction, and difficulty in centralization due to soft tissue tension.[7] It can be applied in patients who have a partial radius and undergo intervention at an early age. However, in the case of the total absence of the radial, the tissues will be stretched more radially. Therefore, there are also authors suggesting centralization after soft tissue distraction.[7,8] Distraction before centralization has some advantages in the treatment of RLD. These include reducing the risk of tissue necrosis by gradual stretching of soft tissues and not requiring carpal bone resection in patients who cannot be centralized in 1 step; as a result, a more stable and mobile wrist can be obtained.[9] In a review study, first distraction of soft tissues and then centralization were recommended to prevent recurrence and reduce complications in RLD treatment.[6] However, some authors have reported that the use of external fixation before centralization causes joint laxity due to stretching in soft tissues, and this leads to recurrence.[10] We also chose soft tissue distraction with an Ilizarov circular external fixator and then centralization in all the patients in our study. All the patients in our series had total absence of the radius. We did not observe skin necrosis or extremity circulation problems in any of our patients. However, as in general external fixator treatments, pin-site infection was observed in our patients. However, we did not have any cases of permanent infection after the fixator treatment was terminated.

Correction of RLD consists of 2 different techniques—radialization and centralization. Radialization provides an advantage of higher ROM. On the other hand, the advantages of centralization are better correction and lower rates of recurrence.[6] In our patients, we achieved soft tissue release with the help of distraction, and we did not expect joint stiffness. We thought that recurrence would be a stiffer problem in our patients. Consequently, we preferred centralization.

In a previous study, the preoperative HFA was 86° on average and 14° in the checkup. The values in our patients were 82.6 and 11.2, respectively, and were similar to those previously reported values. In the same study, the HFP angle was –9° preoperative and +10° in the checkup.[11] In our patient group, these values were similar, i.e., –14.5° and +14°, respectively. In another study, the preoperative UB angle was 61°, while it was 7.3° in our study.[12] In our patient group, the UB angle was less. This may have been because all of our patients had TAR syndrome, and there was no other patient group. Indeed, in that study, the patients had different RLD origins.[12]

Recurrence is a very common condition in patients with RLD.[10,13,14] Due to the fact that the ulna is a single and deformed bone and due to the stretching forces of the muscles attached to the carpal bones, radial deviation is observed again in time. However, recurrence may not always be a significant problem. The high preoperative angle values and the age of the patient determine the recurrence rate.[8,13] In 1 of our patients, there was bilateral recurrence that required an operation.

Although RLD is present in all patients with TAR syndrome, it has been reported in roughly half of the lower extremity patients (47.2%). In other studies, the deformities accompanying TAR syndrome are genu varum, coxa valga, knee flexion contracture, internal tibial torsion, absence of the patella, and absence of cruciate ligaments in the knee.[15] In 2 of our patients, fixed flexion contracture of the bilateral knees, internal tibial torsion, absence of the patella, and laxity (absence of cruciate ligaments) were present. In the literature, the treatment of lower extremity deformities varies from brace treatment to arthroplasty.[16,17] In another study, it was reported that the flexion deformity of the knee was corrected acutely.[16] However, we did not use acute correction because acute correction of a fixed deformity in the knee may cause both circulatory disorder and bleeding. Our treatment method is to open a contracture with an Ilizarov external fixator in the first stage to avoid circulatory disorders, tendon transfer in the second stage to prevent recurrence, and finally to follow the rake with a brace. In the literature in English that we were able to access, we could not find any article mentioning the gradual treatment of flexion contracture accompanying TAR syndrome.

In general, the treatment of fixed contracture deformity in the knee with circular fixators is successful.[18,19] The problems encountered in studies are stress fractures of the femur/tibia during contracture correction, recurrence of the deformity, and pin infection.[18-20] There were no stress fractures in our patients. Although all of our patients had pin-site infection, we did not observe a chronic infection after fixator removal. We increased the preoperative flexion contracture angles from 102.5 (110-95)° to 12.5 (5-20)°; however, some flexion contractures developed again after the treatment in both patients (Table 2). We saw serious knee instability especially in the first months of fixator removal with loosening of the posterior ligaments. To deal with this situation, we kept our first braces rigid. We did not allow knee flexion or extension for the first 6 months. After 6 months, we started to use braces that allow movement in the knee while revising our braces.

Our study has some weaknesses. The number of patients in our study is small. Since TAR syndrome is a very rare condition, it is very difficult to serialize in large numbers in this patient group. The final treatment of patients whose follow-up is started in childhood ends when growth is completed. Our patients continued to grow during the follow-up period.

In the treatment of both lower and upper extremity deformities of TAR syndrome, performing definitive operations following soft tissue distraction with external fixators is an effective and successful method with few complications. However, the use of an external fixator did not prevent recurrence in our series, since 1 of 4 patients had complete recurrence, 3 had partial recurrence, and 1 patient had partial recurrence in the lower extremity. However, we observed that it prevented soft tissue problems in our patients due to the absence of skin necrosis and circulatory disorders. When left alone, these patients did not have any chance of walking because of severe knee

flexion contracture. However, these patients were able to walk, albeit with support, despite some recurrence.

Ethics Committee Approval: Ethical committee approval was received from the Ethics Committee of İzmir University of Economics (Approval No: B.30.2.İEÜSB. 0.05.05-20-130).

Informed Consent: Informed consent was obtained from the legal guardians of the patients before data collection.

Author Contributions: Concept - M.A.; Design - M.A.; Supervision - Ç.B., M.Ö.; Materials - Ç.B.; Data Collection and/or Processing - Ç.B.; Analysis and/or Interpretation - Ç.B.; Literature Review - M.A.; Writing - M.A.; Critical Review - M.Ö.

Declaration of Interests: The authors have no conflicts of interest to declare.

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