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Development and function of a natural reverse shoulder in a patient with thalidomide-induced dysmelia

Michael Kimmeyer, MD [a,*], Lars-Johannes Lehmann, MD [a], Christian Gerhardt, MD [a], Jonas Schmalzl, MD [a,b]

[a] Department of Traumatology and Hand Surgery, St. Vincentius Clinic, ViDia Clinics, Karlsruhe, Germany
[b] Department of Trauma, Hand, Plastic and Reconstructive Surgery, University Hospital Wuerzburg, Würzburg, Germany

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In the late 1950s and early 1960s, the drug thalidomide (Contergan®) was administered to thousands of women across the world due to pregnancy-related nausea. The drug caused severe birth defects which are referred to as thalidomide embryopathy or thalidomide syndrome. Thalidomide survivors were born with a wide range of impairments. Most commonly they have missing or short and/or deformed limbs (phocomelia).[12] This results in different anatomical conditions of the musculoskeletal system which can cause severe physical problems.[8] There are some studies which show that many thalidomide survivors develop osteoarthritis of the upper and lower limb.[3] Globally, thousands of thalidomide survivors and their families continue to live with the medical and social consequences of thalidomide. As they age, they are experiencing new thalidomide-related health problems as well as deterioration in their original impairments. This can lead to disabling and discriminatory outcomes of which we know relatively little. In this case report, a thalidomide survivor with a special form of osteoarthritis is presented.

Case report

In 1961, a male neonate was born with phocomelia of the upper limbs. Since the early years, he had recurrent physical problems concerning both upper extremities and has rarely been completely pain free. Many orthopedic examinations as well as radiological examinations as well as continuous orthopedic treatment were necessary in the course of his life. Figure 1a shows the x-ray examination of both shoulders at the age of 6 months. The left upper limb consists of one long tubular bone and some small, rudimentary bones forming a dysplastic hand. The right upper limb consists of two long tubular bones in addition to a dysplastic hand. The right limb is seen at 1 year 7 months (Fig. 1b). At the age of 4 years and 5 months, the proximal humeral epiphysis with the development of secondary ossifications centers appears (Fig. 1c). The anteroposterior and axial views of the x-ray examination show the right upper limb and the humeral head at the age of 9 years (Fig. 2, a and b). At the age of 21 years, the epiphyseal growth plate is unfused (Fig. 2c).

At follow-up, the 59-year-old man is in good general condition. The highly educated right-handed architect has hardly any restrictions in everyday life and is even able to drive a custom-made car. However, since two years, he has had increasing pain in the right shoulder. Owing to the pain, he takes etoricoxib (a selective cyclooxygenase 2 inhibitor) every two weeks for several days. Interestingly, the left shoulder is completely pain free. Regarding the right shoulder, there were no signs of infection. Palpation showed pressure pain in the area of the bicipital groove and at the insertion of the deltoid muscle. Active range of motion (ROM) is shown in Figure 3. Abduction and flexion were possible up to 180 degrees so that the patient is able to reach the back of his head (Fig. 3e). Internal rotation of the 90-degree abducted shoulder was 30 degrees, external rotation was 80 degrees. The patient was able to perform smooth circular movements in the right shoulder. The isometric abduction and external rotation were painless.

Institutional review board approval was not required for this case report.

* Corresponding author: Michael Kimmeyer, MD, Department of Traumatology and Hand Surgery, St. Vincentius Clinic, ViDia Clinics, Suedendstraße 32, D-76137 Karlsruhe, Germany.
E-mail address: michael.kimmeyer@vincentius-ka.de (M. Kimmeyer).

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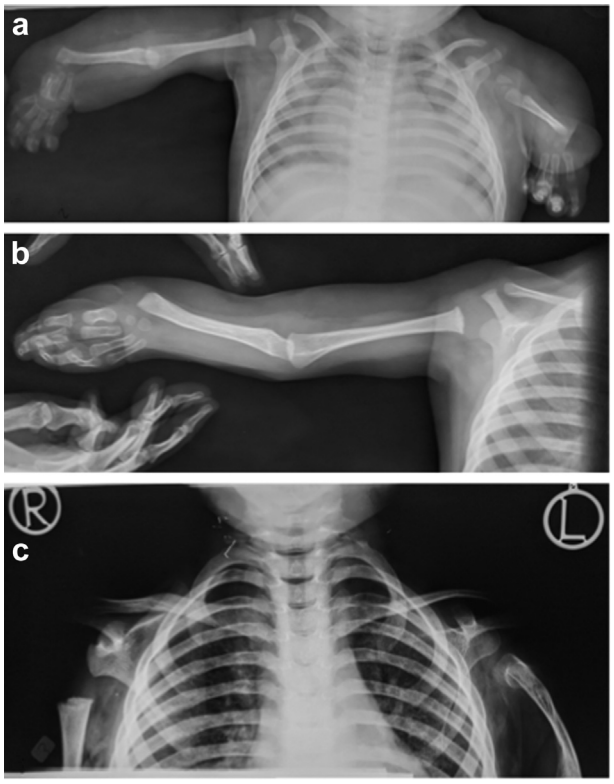

Figure 1 AP view of conventional x-rays. (**a**) Both shoulders at the age of 6 months; (**b**) The right shoulder at the age of 1 year and 7 months; (**c**) Both shoulders at the age of 4 years and 5 months. AP, anteroposterior.

The computed tomographic imaging and the 3-dimensional reconstruction of the right shoulder at the age of 59 years are shown in Figures 5 and 6. The magnetic resonance images of the right shoulder at follow-up are shown in Figure 7. It shows the dysplastic and arthropathic glenohumeral joint with reversed bony glenohumeral structures that is a convex glenoid and a concave humeral head. There are typical osteoarthritic signs, for example, a joint space loss, cystic lesions, osteophyte formations, and a subchondral sclerosis (Figs. 4–7). The rotator cuff shows mild degenerative changes such as for example, an intratendinous lesion of the supraspinatus tendon (Fig. 7b). In conclusion, the patient has a special form of osteoarthrosis with a natural reversed anatomy of the glenohumeral joint of the right shoulder.

Discussion

The drug thalidomide has been approved for sleep disorders and pregnancy-related nausea in Germany since 1957. Taking the drug led to thalidomide embryopathy. The sensitive phase for embryopathy is between the third and sixth week of pregnancy.[10] Typical consequences of ingestion are skeletal deformities. Dysmelia of the upper and lower extremities such as aplasia, hypoplasia, or phocomelia of bones are common. In addition, malformations of the sensory and internal organs as well as brain damage can occur.[13] Throughout life, many thalidomide survivors suffer from thalidomide-related health problems.

The presented patient has hypoplastic upper limbs with only one long tubular bone on the left side. The growth plates of the long tubular bones regularly close in males at around the age of 18 years.[6,11] Consequently, the patient showed a delay in bone development. The development of the patient's glenohumeral joint surprisingly resembles a reverse shoulder replacement which is similar in shape to the shoulder prosthesis invented by Paul

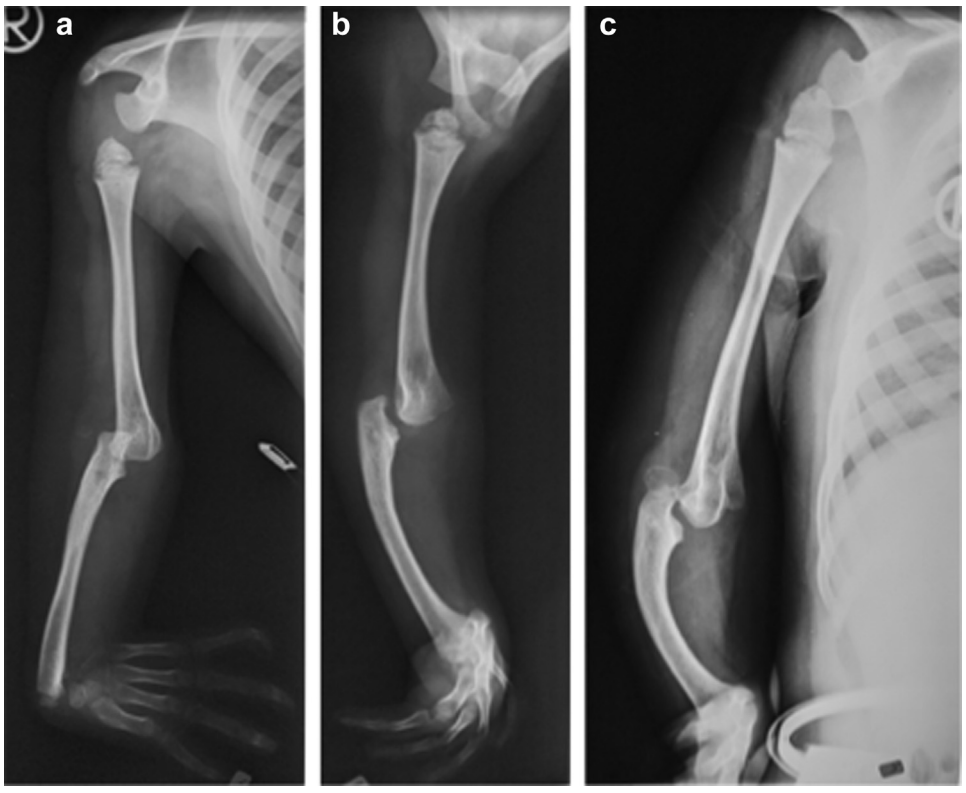

Figure 2 Conventional x-rays of the right shoulder. (**a**) AP view at the age of 9 years; (**b**) Axial . view at the age of 9 years; (**c**) AP view at the age of 21 years. AP, anteroposterior.

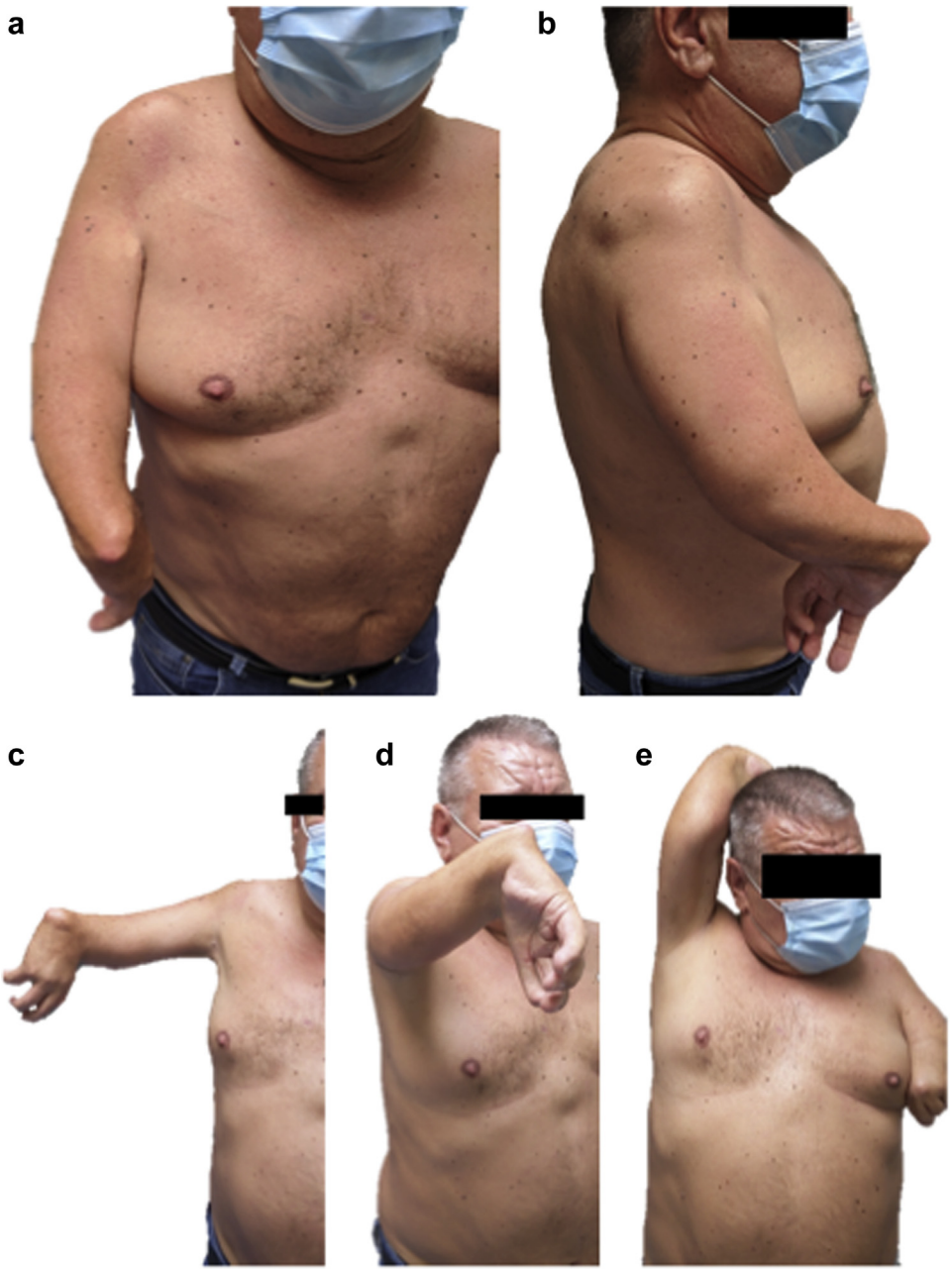

Figure 3 Anatomy and function of the right shoulder at follow-up. (**a**) Anterior view; (**b**) Lateral . view; (**c**) 90° abduction; (**d**) 90° flexion; (**e**) Maximum active abduction.

Grammont in 1985.[4,5] The biomechanical principle of this prosthesis is based on the articulation of a convex glenoid body with a concave joint partner in the humerus. The center of rotation is shifted medially and distally so that the lever arm of the deltoid muscle is extended. This enables a stable center of rotation in the glenohumeral joint for the entire ROM.[1,2]

The analysis of the ROM of the right shoulder of the presented patient shows astonishing adaption processes. The bony, muscular, and ligamentary structures differ significantly from the regular anatomy of the shoulder joint. Nevertheless, the patient is able to perform complex movements so that he has little difficulty in participating in everyday life. There is scarce literature on the long-term consequences of thalidomide embryopathy on the musculoskeletal system. Newbronner et al examined 25 relevant publications in a systematic review, summarized specific health problems, reported on surveys on health and health-related quality of life of thalidomide survivors.[8] Studies on the long-term consequences on the lower extremity have shown that degenerative changes and osteoarthrosis are more common in thalidomide survivors than in the normal population.[3] Newman published the first case report of a patient with thalidomide-induced phocomelia who had a joint replacement.[9] Owing to end-stage glenohumeral osteoarthritis, an anatomical hemiarthroplasty was performed in a 35-year-old woman with good postoperative results. In another case study, Merkle et al described 5 patients who had shoulder replacements due to osteoarthritis. The authors showed improvement of ROM and pain relief in the operated patients.[7]

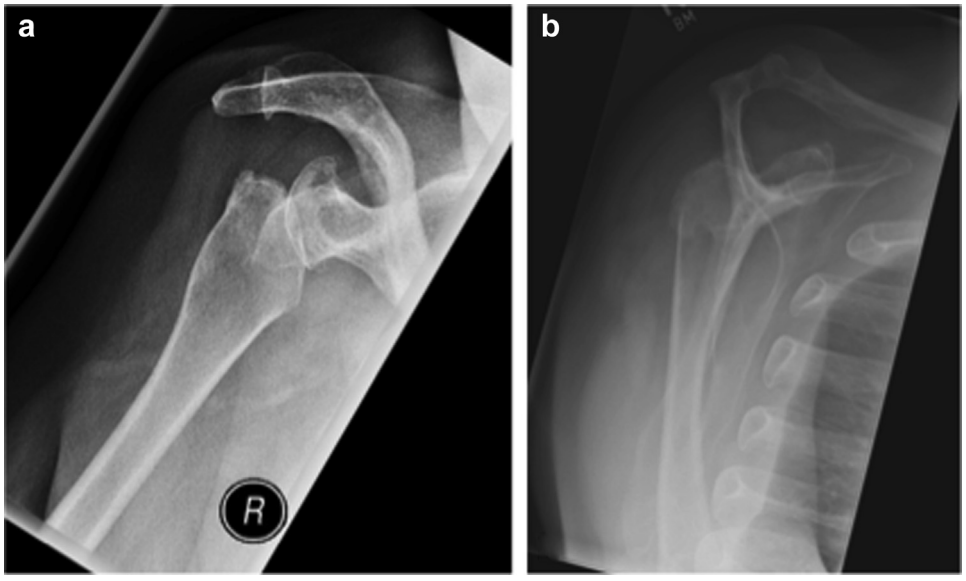

Figure 4 Conventional x-rays of the right shoulder at the age of 58 years. (**a**) AP view; (**b**) Lateral view.

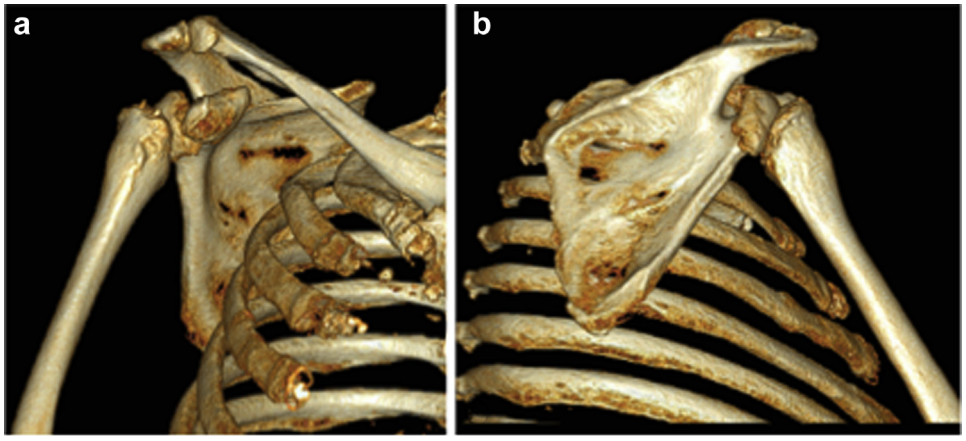

Figure 5 3-dimensional reconstruction of the right shoulder at the age of 59 years. (**a**) View from anterior; (**b**) View from posterior.

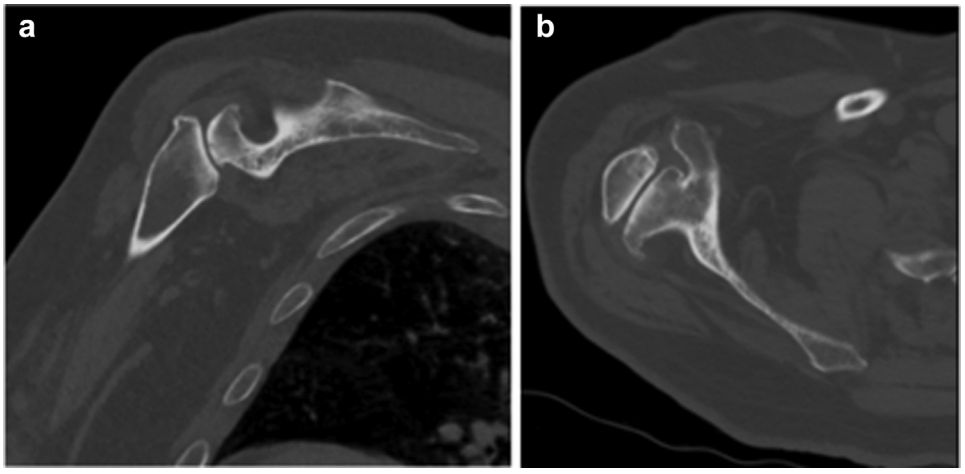

Figure 6 Computed tomographic imaging oft the right shoulder at the age of 59 years. (**a**) Coronal view; (**b**) Axial view.

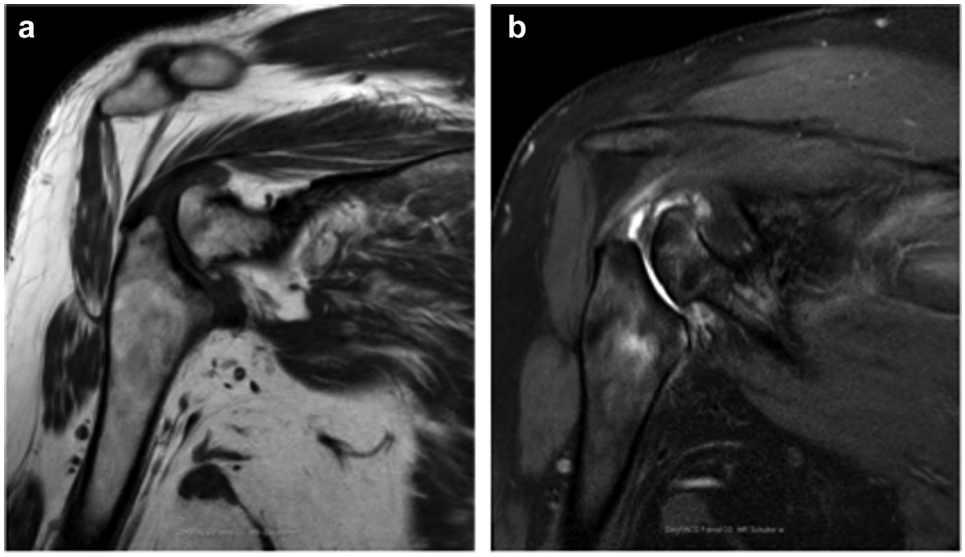

Figure 7 Magnetic resonance imaging of the right shoulder at the age of 59 years. (**a**) Coronal view, T1 sequence; (**b**) Coronal view, PD-TSE-SPAIR sequence.

Conclusion

It is still important to report on the long-term consequences of thalidomide embryopathy to make people aware of the consequences of one of the largest drug catastrophes in history. In addition, the treatment of survivors of thalidomide often requires interdisciplinary collaboration to enable the patient to live a low-complaint life.

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The authors have participated sufficiently in the work to take public responsibility for the entire content of the article.

Conflicts of interest

The authors, their immediate families, and any research foundations with which they are affiliated have not received any financial payments or other benefits from any commercial entity related to the subject of this article.

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Patient consent

Patient consent to publish this report was received.

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