

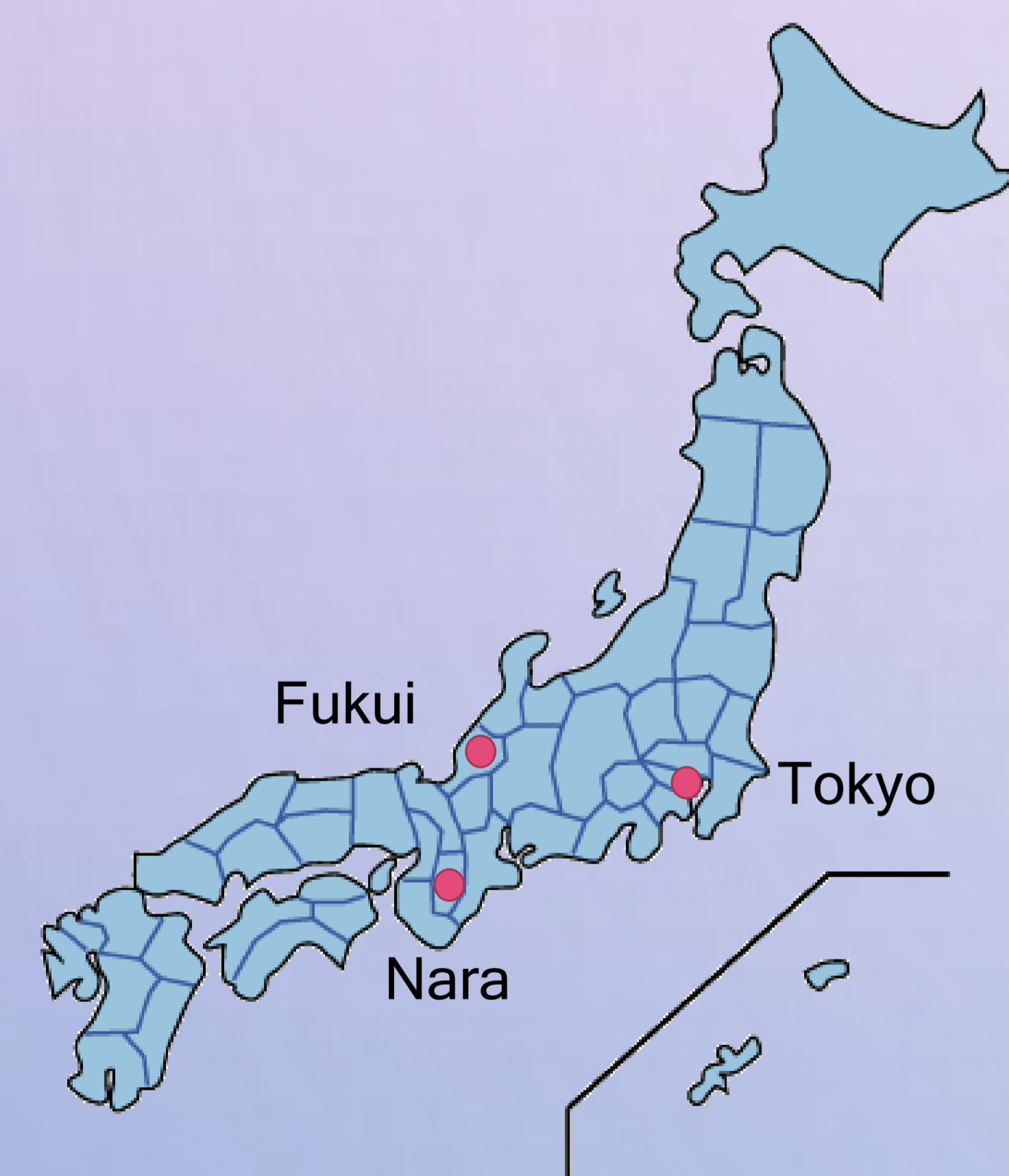
Medical care provided by a private clinic with the highest number of hemophilia patients in the region

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Introduction and Objectives: In Japan, medical care for patients with hemophilia is provided mainly by university hospitals and other general hospitals. However, due to a shortage of specialists in hemophilia care, only a limited number of hemophilia patients may be able to receive specialized care. In our clinic, a single doctor provides comprehensive medical care for all hemophilia patients, which is unusual in Japan.

Methods: After completing a two-year residency at Nara Medical University Hospital, one of the top schools in Japanese hemophilia care, the first author worked in the Fukui National hospital for 8 years and was involved in medical care for both inhibitor-negative and inhibitor-positive and rehabilitation, provision of guidance on self-infusion at home and other aspects of managing hemophilia.

Results: Using the experiences gained during that period, from 3 years ago the first author currently engages in medical practice at a private clinic in the Fukui Prefecture. This Prefecture, has a population of about 78 million people and about 20 people of hemophilia patients. In our clinic provides 9 patients with medical care. It has the highest number of hemophilia patients in the region.



Fukui~Tokyo: about 500km
Fukui~Nara: about 200km

By being closely involved in the treatment of individual hemophilia patients over a long period of time, mutual trust develops, which helps to identify patient concerns and provide better treatment. Achieving this goal requires playing multiple roles in addition to that of a pediatrician specializing in hemophilia; these include being an orthopedic surgeon and also a physical therapist treating arthrosis, a physician focusing on follow-up for lifestyle diseases, a clinical psychotherapist providing psychological care and support, as well as a medical social worker providing consultations on welfare. Needless to say, consultations and referral of patients to a hemophilia specialist in the applicable field when more specialized care is required are also part of the comprehensive management provided at our clinic.

Case: A 29-year-old man with hemophilia A (FVIII: C 2.0%). Complications: Type 2 diabetes mellitus (under follow-up at our facility), attention deficit hyperactivity disorder (ADHD) + depression (receiving outpatient care at a mental clinic). Disease history: Left total hip arthroplasty (THA) for treatment of left femoral head necrosis in 2007 (age 20). Family history: Older brother with hemophilia A (moderately severe).

History of present illness: Because his older brother had hemophilia A, this patient received a coagulation test soon after birth and was diagnosed with hemophilia A. Since then, he had been followed-up at Hospital A. In 2007 (age 20), he opted to receive a checkup by a specialist and was referred to the National Fukui Hospital. At the time of referral, the patient was 169.9 cm tall, weighed 93.1 kg, and was receiving an infusion of factor VIII preparation (2000 U/dose) with the help of his mother upon hemorrhagic events. At that time, the patient was a student of a correspondence course from a senior high school (was only occasionally required to attend school) and was receiving prophylactic replacement therapy (once in 2-3 weeks) at our facility before he attended school. He seldom went outdoors, and the frequency of bleeding was once in 2-3 months (bleeding within the right ankle) or once in 3-4 months (nasal hemorrhage). Before referral to our facility, the patient had never received an articular evaluation. Therefore, during his first visit to our facility, an X-ray image was obtained, and he was diagnosed with right ankle arthritis and left femoral head necrosis. He underwent left THA in the following month. During the hospital stay, he learned to self-inject and began receiving regular replacement therapy with a factor VIII preparation (2000 U × 3 doses/week). He was discharged from the hospital 4 months after admission. He weighed 77.7 kg during discharge. After graduation from senior high school, he did not intend to look for a job because of his depression, and spent his time at home.

Course: In 2013, the author opened a private clinic, and the patient began to receive care at this clinic (age 26). In those days, the patient's parents divorced and the patient began to live with his father and older brother. His father retired from his job owing to the age limit, and his older brother remained jobless for many years because of depression. His father spent the retirement allowance and began to borrow money for daily living from consumer loan companies. At the same time, the father began a part-time job. His older brother became aware of the crisis and began to work at a part-time job as well. The patient, after realizing the situation, applied for a part-time job in response to various recruitment announcements and was finally employed at a shop. At that shop, however, he was unable to do the work like his colleagues because he had ADHD and was reprimanded by the manager frequently. This caused him stress, resulting in melena every day. Because he had depression, he spent most of his energy working as a part-timer and mostly remained lying on the bed while at home. He often bought food at nearby convenience stores and seldom went outdoors, thus making it difficult for him to reduce his body weight. During each visit to our clinic, the author listened to what the patient reported, and occasionally, advised him about daily living. One year later, the patient was forced to quit the job at the shop.

Fortunately, he found a new part-time job position soon and is now working there. However, even at the new place of work, he is often reprimanded by the manager during work. In addition, he sometimes remembers his mother. For these reasons, he has attempted suicide by cutting his wrist. He commutes to the place of work on bicycle (on foot in winter because of snow). Because he has ADHD, he often collides with the roadside signboards, falling from the bicycle onto the road or into roadside gutters. Every time such an event occurs, he receives an infusion (2000 U/dose). As a result, the regular replacement therapy, initially planned for 3 times a week, has been actually applied 4-5 times a week. Furthermore, he often sustained sprains while running or walking on flat roads during work. For this reason, the author advised him to buy trekking shoes and increased the dose level to 3000 U/dose on Tuesdays and Thursdays and to 4000 U/dose on Saturdays (FVIII following 3000 U infusion: C trough level 9%, peak level 85%).

After the dose increase, the frequency of bleeding decreased but has not reached zero. Despite the advice to practice voluntary rehabilitation, he seldom practices rehabilitation because he spends most of his energy at work. During each visit to our clinic, the author listens to what the patient reports, provides rehabilitation, and recommends him to pursue the procedure for receiving a pension for the disabled.

Conclusions: When a patient is approached by a single doctor focusing on various aspects of the disease, both medical and social improvement in the patient's quality of life can be achieved.



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