Q&A on Thalidomide-Impaired People

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This Q&A was put together for doctors, nurses and other healthcare professionals, who need to examine and treat thalidomide-impaired people but are not fully familiar with thalidomide impairment, so please introduce it to doctors and other staff as necessary. This booklet will also be immediately useful for doctors who only have time to read the relevant sections.

We hope that this booklet will help in maintaining health, preventing disease and providing routine medical care for thalidomide-impaired people around the world, over half a century after the deleterious effects of thalidomide first surfaced.

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Recommendations and Suggestions

Recommendations

１、It is strongly recommended that health care workers utilize this Q&A.

２、It is recommended to use automated sphygmomanometer in cuff-oscillometric methods on posterior tibial artery for measuring blood pressure for people with upper limb disabilities.

・Measurement should be done in recumbent position.

・When the normal (size M) cuff is used to measure in the method stated above, systolic pressure should be (posterior tibial artery systolic pressure＋8)×0.88 mmHg

・It is recommend to measure blood pressure at home.

・When arm blood pressure is measured, it is strongly recommended to use cuff that is suited for the upper arm circumference.

３、If a person with upper limb disabilities shows Sokolow-Lyon index SV1＋RV5≧3.5mV or R5 or R6≧2.6mV in the electrocardiogram, left ventricular hypertrophy is suspected so it is strongly recommended to do an echocardiography.

・There is a possibility that potential patients with high blood pressure is found.

４、If it is difficult to take blood because of the disability of upper limb, evaluate whether they have fatty liver with abdominal ultrasound.

・If fatty liver is found, there is a high possibility that they have lipid metabolism abnormality so blood chemistry study is strongly recommended.

５、If they feel pain in forearms and upper arms because of carpal tunnel syndrome, it is strongly recommended to evaluate their cervical vertebrae.

・There is a possibility that proximal symptom is a sign for cervical spondylosis.

６、It is recommended that upper gastrointestinal endoscopy is done via nasal endoscope by a trained doctor.

・There is less pain for the patient and sedative is not necessary.

７、It is strongly recommended to not wear a mask when examining a patient with hearing impairment.

・Patients with hearing impairment naturally acquire skills (lip reading) to visually supplement the lack of hearing.
・A doctor should not talk to patients with hearing impairment from the angle where they cannot see the doctor's mouth, such as facing the electronic chart.

８、It is recommended to use explanation material indicated in this Q&A when examining patient with hearing impairment.
・There may be a necessity for communication by writing so prepare a whiteboard or memo pad.

９、It is recommended to do health check for 2 people with hearing impairment on the same day.
・They can share a sign language interpreter.
・They feel a sense of security because patients with hearing impairment can communicate with each other.

10、It is recommended to consider whether they have mental disorder such as depression.
・The results of the survey of actual life situation showed that disease rate is higher (10.4%) than the people in the same generation (2.0%).

Proposals for the way comprehensive assistance to the Thalidomide-impaired people should be

1. To propose to construct the system for providing information and consultation in order to promote the positive use of the system to assist the life of Thalidomide-impaired people.

2. To propose to continuously assist the Thalidomide-impaired people to receive a regular health check because they are more likely to suffer from the lifestyle diseases than general public.

3. To propose to foster doctors and promote medical institutions with whom/which the Thalidomide-impaired people and their doctors in charge can consult about

the treatment principles related to “the pain” and sequelae.

4. To propose to promote international association so that the Thalidomide-impaired people and medical staffs can share the knowledge that has been gained as a result of studies done in Britain, Germany and Japan.

5. To propose to ensure that the medical students study the history of drug disasters and prevention as well as the Relief System for Sufferers from Adverse Drug Reactions.

Continuance of the studies

This study is still continued even after April 2014. The new representative is Fumihiko Hinoshita, Nephrology, National Center of Global Health and Medicine

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13. Source materials

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Source 2: Hearing-impaired patients: Upper gastrointestinal endoscopy (through the mouth) Supporting document

Source 3: Hearing-impaired patients: Upper gastrointestinal endoscopy (through the nose) Supporting document

Source 4: Hearing-impaired patients: Respiratory function tests supporting document

Source 5: Hearing-impaired patients: Eye tests supporting document

Source 6: Hearing-impaired patients: Gynecological examination supporting document

1． Basic facts about thalidomide impairment

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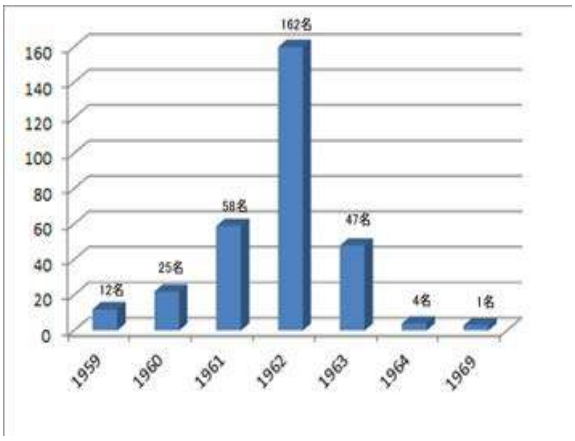
Q1-1-4: Ryoji Kayamori
Q1-5: Misato Tanaka
Q1-6: Ryoji Kayamori
Q1-7: Atsuto Yoshizawa

Q1-1: When did the thalidomide crisis occur and how many Japanese victims are there?

・309 people were recognized as being affected by thalidomide in Japan, and 295 of these were still alive as of April 2012.

Thalidomide was first marketed in West Germany in 1957 as a sleep medication under the name Contergan. It went on sale in Japan in 1958 as the sleep medication Isomin. In 1960, the digestive medicine Pro-ban M, which contained a small amount of thalidomide, also came on the market. Infants with thalidomide-induced defects were born to mothers who had taken thalidomide in early pregnancy. Birth defects were observed commencing in 1959 and reached a peak in 1962 (Fig. 1). Three hundred and nine people were recognized as being affected by thalidomide in Japan, 295 of whom were still alive in April 2012. The total number of victims worldwide is estimated at 5,850.

Fig. 1 Number of infants born with thalidomide-impairment in Japan



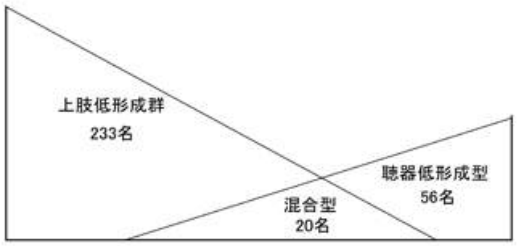
Q1-2: What types of birth defects did thalidomide cause?

There are three patterns of thalidomide-induced defects: victims either have upper limb reduction defects (also called upper limb phocomelia, upper limb underdevelopment, etc.), hearing organ

reduction defects or a mixture of the two.

Upper limb reduction accounts for 75% of the defects, with the remaining 25% being hearing organ reduction and mixed defects.

Fig. 2　Number of Japanese thalidomide victims with each defect type



上肢低形成群：Upper limb reduction defects: 233
混合型：Mixed: 20
聴器低形成型：hearing organ reduction defects: 56

Q1-3: What are the features of upper limb reduction defects?

- These defects range from absence of upper limbs to underdevelopment of the thenar muscles (the group of muscles in the palm at the base of the thumb) and a triphalangeal thumb, and include defects between these two extremes.

Three Japanese victims also have underdeveloped lower limbs, one of whom relies on a wheelchair for mobility because of the severity of underdevelopment.

Q1-4: I didn’t know there were thalidomide-impaired patients with hearing organ reduction defects. What are the features of hearing organ reduction defects?
These defects are mainly accompanied by sensorineural or mixed hearing loss.

In many cases, there is also absence or underdevelopment of the abducens nucleus, facial nerve nucleus or nerves peripheral to these. The oculomotor nerve compensates for the absence or underdevelopment of the abducens nucleus or nerve, resulting in Duane's syndrome. Facial nerve paralysis and Bogorad's syndrome are also common complications.

Q1-5: What are the important points when examining patients with hearing impairment?

- **Both the patient and the doctor should make sure that the speaker's face is visible. Talking while wearing a mask must be avoided, as must talking while facing a computer.**

Hearing impairment (hearing loss) is often difficult to appreciate because it is an invisible impairment, and many people find communication a challenge even if (or perhaps because) their hearing loss is mild.
Communication difficulties arising from impaired hearing, and the responses to those difficulties, differ according to the extent and nature of the hearing loss.

1. People with mild hearing loss

People who can hold a conversation and have no obvious pronunciation problems do not give the impression of having hearing loss. From the standpoint of the person with hearing loss, this can be a problem if, for example, the other person is soft-spoken, lacks clear pronunciation, speaks quickly, or if there is extraneous noise, etc. In such situations, the person with mild hearing loss may mishear or may miss some of what is said, and will naturally ask the speaker to repeat what they have said. If this happens, it is important to ask if the person has hearing loss.

People with impaired hearing naturally learn to visually compensate for their hearing deficit through lip-reading, irrespective of the severity of hearing loss. When talking to a hearing-impaired person, it is therefore necessary for both parties to ensure that the speaker's entire face is visible. Talking whilst wearing a mask must be avoided, as must talking while facing a computer.

2. Hearing aid users

Most people use hearing aids when their hearing loss becomes moderate or worse. However, hearing aid users are still unable to hear like ordinary people. Conversation is difficult in groups, in noisy environments, when the listener and speaker are at a distance from each other, etc.

3. Severely hearing-impaired and profoundly deaf

As hearing loss becomes more severe, hearing aids become less useful. For these people, sign language is an important means of communication. A sign language interpreter will often accompany the patient to the hospital, but written communication must be used if no interpreter is available. A pen and paper should be prepared for such situations.

Whether through lip reading, sign language or writing, the hearing impaired person obviously relies on vision for communication. If tests need to be carried out under dark conditions, strategies are needed to deal with this.

Q1-6 Are there any impairments other than in the upper limbs and hearing organs?

- Other impairments can include organ malformations, such as congenital absence of the gallbladder, heart malformations, mental handicaps, scoliosis, spina bifida occulta, block vertebrae of the cervical spine, lumbarization of the sacrum (L6), hip dislocation due to hip dysplasia, etc.

Q1-7: Where can I find medical literature on thalidomide?

- **There are no websites with collections of medical articles on thalidomide.**

This Q&A was prepared because there is currently nowhere that healthcare professionals can turn to when faced with questions about thalidomide in routine medical practice.

The following websites may provide useful information.
(Japanese and English language sites only)

- http://www008.upp.so-net.ne.jp/ishizue/ Japan
- http://www.thalidomideuk.com/ United Kingdom
- http://www.thalidomidesociety.co.uk/ United Kingdom
- http://www.thalidomide.ca/summary/ Canada
- http://www.thalidomide.ca/links/ Canada

2. Comments from the Department of Orthopedic Surgery and Department of Rehabilitation

Main author: Ryoji kayamori

Q2-1: What are the causes of shoulder pain?

- **There are several contributory factors, the main ones being underdevelopment of the humerus and the humeral head that form part of the shoulder joint. Atrophy of the shoulder girdle muscles and spondylosis are other possible causes.**

1) Underdevelopment of the shoulder joint

Underdevelopment of both shoulder joints is seen in over 100 Japanese thalidomide victims. In Fig. 1, there is underdevelopment of both the left and right humerus, with dislocation of the right shoulder. This is a result of underdevelopment of the shoulder girdle muscles rather than any problems inside the joint.

Advice: In day-to-day life, shoulder pain can be managed by avoiding holding things with the hands, etc. Conservative physical therapy is appropriate, and heat therapy and massage will be effective to a certain extent. Endoscopic therapy is unsuitable.

Fig. 1 Dislocation of the shoulder joint

2) Underdevelopment of shoulder girdle muscles

If there is underdevelopment of the humerus or the shoulder joint is dislocated, the corresponding shoulder girdle muscles will also be underdeveloped and become easily fatigued due to the weight of the upper limb or when carrying bags etc. (Fig. 2).

Advice: Overuse of the underdeveloped muscles should be avoided. Treatments such as heat therapy and massage are appropriate.

Fig. 2 Atrophy of shoulder girdle muscles

3) Spondylosis

The cervical vertebrae progressively degenerate with age. Figure 3 shows an X-ray of a 40-year-old, in which C4/C5 are already unstable under cervical flexion.

Advice: Heat therapy and massage are appropriate for the shoulder pain. Patients with upper limb numbness, muscle weakness in the hands and fingers and marked cervical instability will need to wear a neck brace. We use cervical collars, which are easy to fit.

Fig. 3 Cervical instability

4) Block vertebrae

When block vertebrae develop in the cervical spine (Fig. 4), the range of movement at the block is limited, resulting in excessive loading on the surrounding vertebral bodies and more rapid worsening of spondylosis.

Advice: The first line of treatment is conservative symptomatic treatment of shoulder stiffness.

Fig. 4　Cervical block vertebra

Cervical spine X-rays demonstrating the formation of blocks at C2−C4 and C5−C7.

Upper image: 1976 (Teikyo University Hospital)

Lower image: 2012 (National Center for Global Health and Medicine)

5) Poor posture/kyphosis (hunchback)

Patients tend to bend their trunk forward to compensate for the impaired ability to reach for something **(to stretch out one's upper limbs to reach for something but cannot reach)** due to shortened upper limbs. This brings the head in front of the line of gravity, which places a considerable burden on the neck and shoulder girdle muscles (Fig. 5).

Advice: The posture should be corrected by occasionally lying on the back and placing the back of the head on the floor. Stretching exercises are also effective. The most important thing is to develop good lifestyle habits, such as avoiding overwork and stress, taking walks, etc.

Fig. 5 Kyphosis

The X-ray demonstrates development of kyphosis in the thoracic spine, together with pronounced lumbar lordosis. Shoulder stiffness arises because the head is positioned forward of the body's line of gravity. Mild scoliosis is also evident.

6) Droopy shoulders

Patients with 'droopy' shoulders (Fig. 6) are prone to develop shoulder stiffness when carrying bags or backpacks, as this stretches or compresses the brachial plexus.

Advice: Patients should avoid carrying baggage or heavy objects in their day-to-day life.

Fig. 6 Droopy shoulder

Swan neck and droopy shoulders down to the 1st thoracic spine can be seen in the lateral view of the X-ray of the cervical spine. C4/C5 instability is also evident.

Q2-2: What causes low back pain?

- **Low back pain involving chronic muscle fatigue is becoming more common.**

Low back pain is divided into three categories, being either caused by metastatic cancer of the bones, pathologic fracture, infection, etc., associated with sciatica or sensory disturbance of the lower limbs, or involving chronic muscle fatigue. Low back pain involving chronic muscle fatigue is becoming more common among thalidomide victims. However, in some patients, it is associated with discitis. It may therefore be necessary to use diagnostic imaging such as MRI, if needed, to identify the cause of the pain.

Stress and overtiredness should be avoided, because low back pain involving muscle fatigue can be stress-related. Less common factors, such as scoliosis-related poor posture, spina bifida occulta and lumbarization of the sacrum can also be contributory factors.

1) Scoliosis

Scoliosis is a frequent complication in thalidomide-impaired patients, particularly scoliosis of the thoracic and lumbar spine. Some patients also have spina bifida occulta at L5 (Figs. 7, 8).

Fig. 7 Scoliosis

Fig. 8 Scoliosis and spina bifida occulta

The lumbar vertebrae are convoluted and spina bifida occulta is present at L5.

2) Discitis

An MRI scan is necessary if low back pain is chronic and refractory. The patient is sometimes incidentally found to have intervertebral disc herniation and inflammation (Fig. 9).

Fig. 9 Discitis

The MRI scan shows degeneration of the intervertebral discs, with partial destruction of L2 and L3. Post-inflammatory soft tissue can be seen in the spinous process of the lower back.

Q2-3: What are the main causes of hand numbness?

Skeletal underdevelopment of the hand joints is accompanied by narrowing of the carpal tunnel, so that overuse of the hands leads to carpal tunnel syndrome.

Until now, 12 Japanese thalidomide victims have undergone surgery for this condition, but the number of thalidomide victims affected by carpal tunnel syndrome is probably much higher, because many have not undergone detailed examination despite having hand numbness. This condition is seen in people with mild to moderate upper limb reduction defects (Fig. 10), of which there are about 130 Japanese patients.

Treatment methods: Firstly, the cause of numbness must be investigated. Nerve conduction in the upper limbs is studied by electrophysiological methods. Motor nerve conduction tests of the median nerve are difficult because of absence of the thumb or underdevelopment of the thenar eminence. The presence of a conduction block and axonal degeneration are investigated by inducing a sensory nerve action potential (SNAP) in the index finger via recording electrodes (Fig. 11). Intraoperative findings show that the median nerve is comparatively large because of the narrowness of the bones forming the carpal tunnel (Fig. 12). The distal end of the transverse carpal ligament distal to the constricted part corresponds to the site of abnormal conduction.

Fig. 10 Absence or underdevelopment of the thumb

The left hand presents symptoms of carpal tunnel syndrome due to overuse.

Fig. 11 Electrophysiological study findings

There is conduction block and conduction delay at the hand joint articulations in both hands, suggesting carpal tunnel syndrome. Conduction disturbance in the left hand joint articulations improved after transverse carpal ligament release surgery in this hand, which was more severely affected.

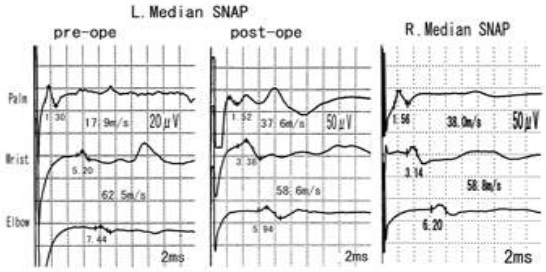
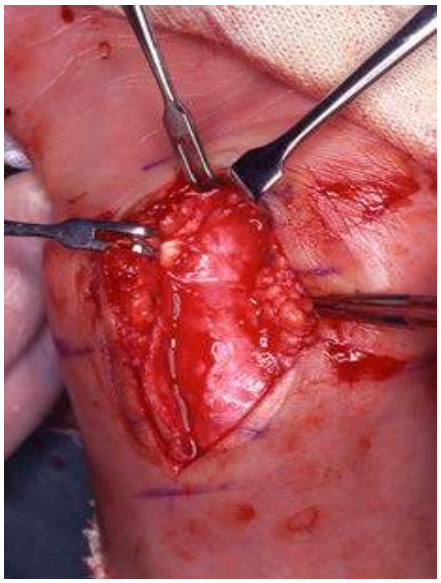


Fig. 12　Intraoperative findings

The median nerve is relatively large, and hence, is easily mistaken for the flexor tendons of the hand joints, including the long palmar muscle. Care must also be taken to avoid cutting the recurrent branch of the medial nerve when its presence within the operative field is not clear.



Q2-4: What causes persistent pain in the hands?

When there is underdevelopment of the musculoskeletal system, activities using the hands can quickly cause fatigue in the forearm muscles and intrinsic muscles of the hand, leading to tendonitis.

Conditions that were observed, in order of decreasing frequency, included de Quervain's disease (tendonitis of the radial and dorsal sides of the thumb), trigger finger, lateral epicondylitis, medial epicondylitis, etc.

Advice: These conditions are hard to cure because it is difficult to restrict the use of the hands in daily life. Refraining from using the hands is the most effective therapeutic approach. Local corticosteroid injection into the tendon is another option, but the problem will immediately recur if the hands are routinely overused again.

Q2-5: What are the causes of hip joint pain?

- **Patients with hip joint pain should be X-rayed to check for degenerative hip disease (coxarthrosis).**

Only 2 of the 309 Japanese thalidomide victims have clear underdevelopment of the lower limbs. However, a number of patients have hip joint dislocation and late-onset coxarthrosis associated with underdevelopment of the acetabulum. Hence, patients with hip joint pain should be X-rayed to check for coxarthrosis (Fig. 13).

Coxarthrosis becomes progressively worse with age, because the hip joints support the weight of the body (Fig. 14). Hip replacement surgery will remove the pain and improve the patient's activities of daily living. However, a number of thalidomide victims with upper limb reduction defects use their lower limbs to perform upper limb functions (Fig. 15). After hip replacement, these patients are therefore highly prone to dislocation of the artificial hip through adduction of the lower limbs.

Advice: Patients should thoroughly discuss their lower limb use with the orthopedic surgeon to determine whether surgical treatment is appropriate.

Fig. 13 Underdevelopment of the hip joint

X-ray of the lumbar spine and hip joints of a teenage patient. Both hips already display mild coxarthrosis.

Fig. 14 Coxarthrosis

Left coxarthrosis has steadily deteriorated and the hip is dislocated. The patient would be indicated for hip replacement surgery.

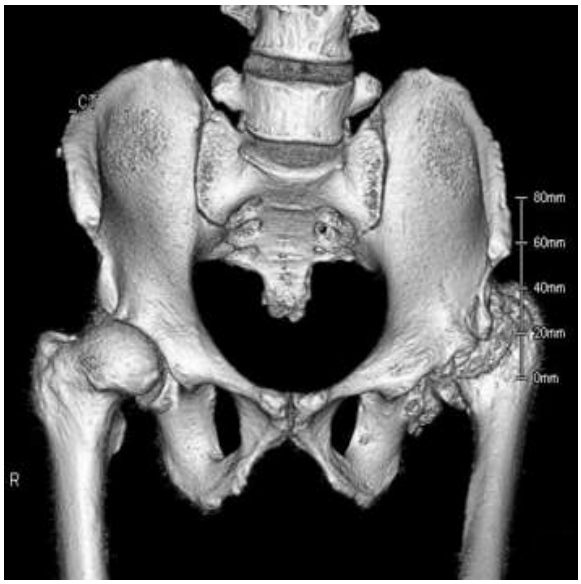
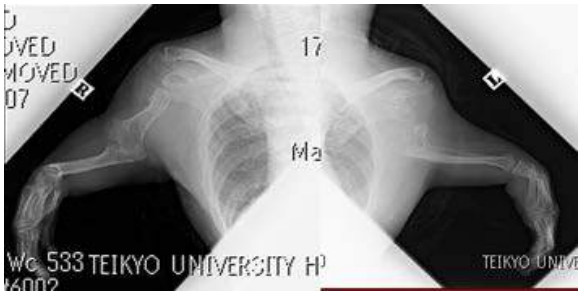


Fig. 15　Upper limb reduction defect

The lower limbs have been used to compensate for the upper limb reduction defects. In such cases, hip replacement surgery would not be appropriate.



3. Comments from radiologists

Main authors

Q3-1: Kazuyoshi Yamano, Toru Sasaki, Tatsuya Wada, Kanehiro Hasuo

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Q3-1: What are the important points when taking X-rays?

- **Patients have different degrees of impairment, but regardless of the degree, for patients with upper limb reduction defects in particular, care should be taken that they do not fall from the X-ray table or fall over when moving, because, if they lose their balance, they cannot use their upper limbs to support themselves.**

Many thalidomide-impaired people have had to undergo a great number of examinations at medical institutions from a young age. Some will therefore feel uneasy and more fearful or resistant than ordinary patients when attending medical centers, and should be treated with care and kindness in response to these fears. Care should be taken to not place too much burden on the patient when he/she is adopting the positions needed for X-ray, as this can easily damage the relationship of trust with the radiologist. Patients range from those with relatively mild impairments, who live independently and will require almost no assistance in changing their position, raising their arms, etc., to those with severe impairments who require help with most things, such as getting undressed and dressed, etc. However, whatever the extent of impairment, care should be taken that the patient does not fall from the X-ray table or fall over when moving, because if they lose their balance they cannot use their upper limbs to support themselves. Below, we describe the important points to remember when taking X-rays of the chest (AP and lateral), cervical, thoracic and lumbar spine in two planes, both shoulders (AP and Y view), hip joint (AP), both knees in two planes, etc., and when X-raying patients with impaired hearing.

3. Comments from radiologists

Chest X-ray

(Fig.1)　Chest x-ray (AP)

(Fig. 2)　Chest x-ray (lateral)

Chest AP image (Fig. 1)

The patient stands and places their chest against the X-ray plate.

The patient is asked to hold their arms out in front.

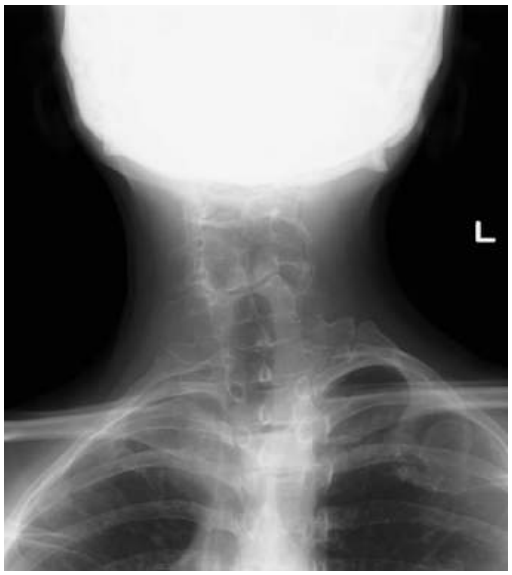
Chest lateral image (Fig. 2)

After the AP X-ray, the patient is asked to turn sideways and, if possible, to raise their arms as high as possible. Patients who cannot raise their arms by themselves need assistance.

If the arms are too short to be raised even with assistance, the patient is asked to hold their arms out in front as far as possible.

Cervical spine X-ray

The cervical spine can be X-rayed with the patient seated. This requires no movement.



(Fig. 3) Cervical spine AP

(Fig. 4) Lateral

Cervical spine AP (Fig. 3)

If the cervical spine is deformed (Fig. 3), the X-ray can be taken with the patient's head tilted slightly up and down, in turn.

Cervical spine lateral (Fig. 4)

The patient should be seated sideways with the back straightened and the gaze fixed on a single point (Fig.4).

Help should be given to support the head if images with the head bent forwards and backwards are needed.

Thoracic and lumbar spine

For X-rays in the decubitus position, assistance in the form of back support should be given even if the patient can adopt the position on their own. When changing from decubitus to lateral decubitus positions, the patient should be allowed to move on their own, but with staff remaining at their side and providing assistance if necessary. Care also needs to be taken when patients with shoulder malformations adopt the lateral decubitus position, as this is painful.

3. Comments from radiologists

Thoracic spine AP

(Fig. 5)

Thoracic spine lateral

(Fig. 6)

Lumbar spine AP

(Fig. 7)

Lumbar spine lateral

(Fig. 8)

Hip joints AP

This requires inward rotation of both lower limbs, but should not be forced if it is painful. Instead, the patient should be asked to turn the lower limb inwards as far as possible on their own.

(Fig. 9)

Shoulder joints

Shoulder joint true AP (Fig. 10)

Shoulder joint scapular Y (Fig. 11)

Shoulder joint true AP (Fig. 10)

This is taken in the same position that allows visualization of the glenohumeral joint and subacromial joint in normal individuals.

Here, the patient was seated in front of the X-ray plate and the image was taken with the target shoulder at an angle of 30 degrees and the X-ray center line at 20 degrees to the craniocaudal plane.

The subacromial joint could not be captured because of deformation of the scapula.

Shoulder joint scapular Y (Fig. 11)

This is taken with the coronal plane at 70 degrees to the X-ray plate, to separate the scapular body from the ribs.

Here, the image was taken with the X-ray center line at 20 degrees to the craniocaudal plane.

3. Comments from radiologists

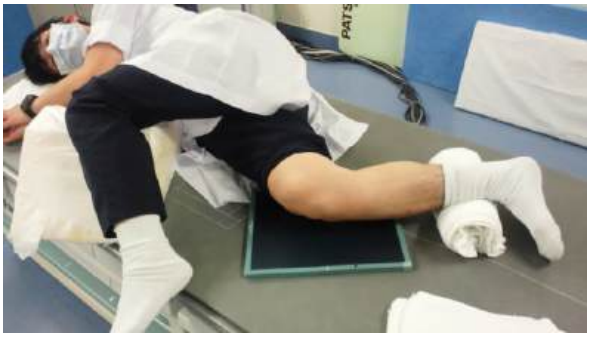
Knee joints

AP view

The X-ray is taken with the patient in the supine position, with the knee extended and the patella at the center.

Lateral view

The patient adopts the lateral decubitus position on the side of the lower limb of interest, the pelvis is placed in the true lateral position, and the lower limb of interest is bent at 30 degrees and held in place by supporting the foot with a pillow, rolled up towel, etc. The other lower limb is supported with a support block and the X-ray is taken. (Fig. 12)



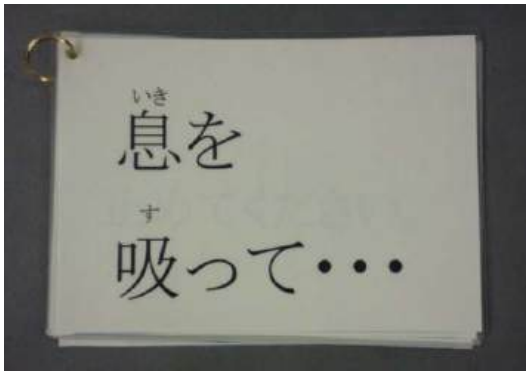
(Fig. 12)

(Fig. 13)

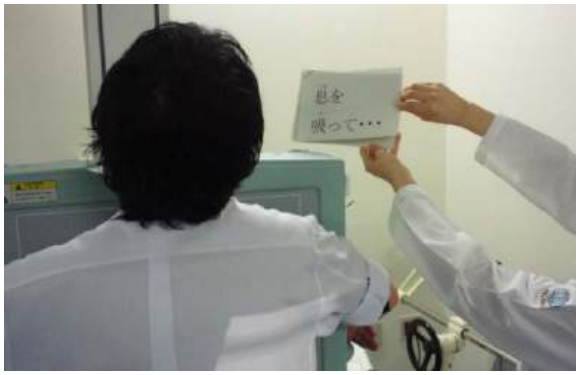
If adopting the lateral decubitus position is difficult, an X-ray can be taken from the side, with the patient in the supine position and the knee of interest supported from below by a support block. (Fig. 13)

Patients with hearing impairments

Instruction cards should be prepared before taking X-rays, and the patient should be asked to follow the instructions shown on each card. (Fig. 14), (Fig. 15)



(Fig. 14)



(Fig. 15)

Q3-2: What points should be considered when receiving thalidomide-impaired patients for X-ray?

- **Patients with upper limb reduction defects have difficulty putting on and taking off examination clothing that needs to be tied at the front, and will therefore need a one-piece examination gown.**
- **Hearing impaired patients will need to be given instructions in writing, or through gestures, lip reading, etc.**

  Most hearing-impaired patients use lip-reading, so you should remove your mask and speak slowly.

① What preparations should be made before X-ray?

The radiologist in charge of the X-ray procedure should check the medical questionnaire attached to the patient's medical chart, and should gain an understanding of the patient's degree of impairment and independence, current subjective symptoms, etc. It is also important to closely monitor the patient's physical and mental state throughout the tests by communicating with the patient. This will enable the radiologist to predict any somatic effects and to prevent accidents, such as falls, when taking X-rays.

If further radiology tests are to be performed, it is important to share physical information on the patient with other departments in preparation for future tests. This will make things easier for those performing the tests and will also give the patient a sense of security.

② What kind of clothing is worn for X-ray tests?

Many imaging tests in the radiology department require the patient to change out of their clothes and into examination clothing. This is difficult for some patients, so every patient should be asked before the examination if they need assistance in changing.

Two types of clothing are used at our hospital: a pajama-type set with a jacket that is tied at the front, and a one-piece gown that is put on over the head.

In order to ensure ease of changing clothes, preparations should be made in advance to enable patients to select the type of examination clothing.

Many patients with upper limb reduction defects find it difficult to wear the pajama jacket as this must be tied at the front. A one-piece gown should be provided for such patients.

③ Please discuss the body positions during X-ray.

Some patients complain of pain when undergoing tests and scans while lying down. It is, therefore, important that patients undergo imaging in as comfortable a position as possible by, for example, adjusting pillow height, placing cushions or towels beneath the feet, etc.

3. Comments from radiologists

During lateral chest X-rays, it is very difficult for patients with shortened upper limbs to raise their upper limbs on their own. They should be given help by providing support for both their upper limbs or getting them to hold an IV stand, for example. (Fig 1)

All patients with upper limb reduction defects, regardless of the degree of impairment, can find it painful to maintain the same position when in the decubitus or lateral decubitus position, and should be given help and support according to their needs.

The radiologist must treat such patients with great care and thoughtfulness, as too much distress during X-ray can easily damage the patient's trust in the medical staff.

(Fig. 1)

Hearing-impaired patients

① Preparations before X-ray

Patient information should be checked before the tests, in the same way as for patients with upper limb reduction defects.

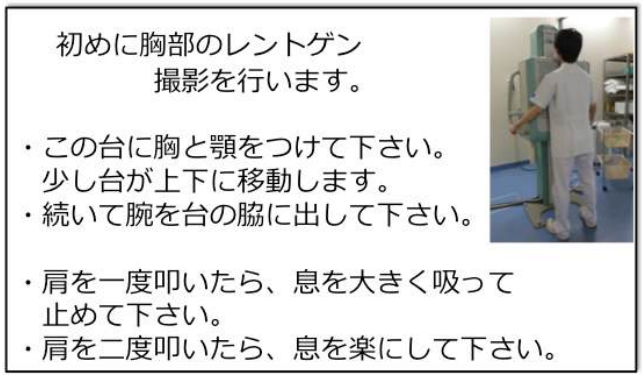
If the patient is accompanied by a sign-language interpreter, the overall test procedure and specific details of the test should be explained through the interpreter before entering the imaging room. The patients should also be asked if they have any questions.

② Points to remember during tests

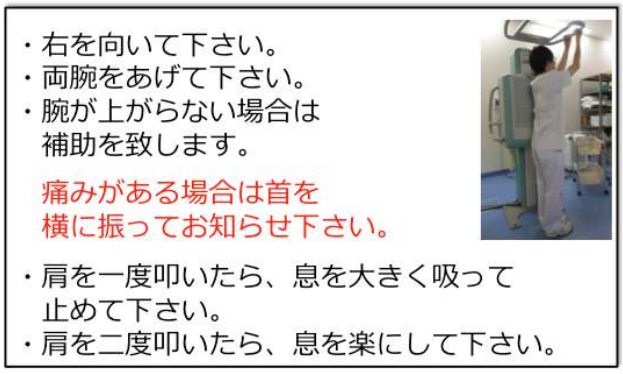
Instructions should be conveyed in writing or through gestures and lip reading in the imaging room. Most hearing-impaired patients use lip-reading, so you should remove your mask and speak slowly.

The test procedure and specific instructions can be easily explained to the patient using cards etc. (Fig. 2.3). The use of photographs or illustrations is also effective in showing the patient what the test will be like.

Distress can quickly be alleviated if the radiologist gets the patient to give an obvious signal when in pain or discomfort during the test.



(Fig 2)



(Fig 3)

Q3-3: What points should be considered when measuring bone mineral density?

- **The lumbar spine and proximal femur need to be measured.**
- **13% of male patients have osteoporosis based on lumbar spine YAM (PR).**

(YAM: Young Adult Mean, PR: Peak Reference)

We compared lumbar spine YAM (PR), femoral neck YAM (PR) and total hip YAM (PR) in 44 thalidomide-impaired patients.

Osteoporosis was found in 13% of men and 4% of women based on lumbar spine YAM (PR), 6% of men and 14% of women by femoral neck YAM (PR) and 0% of men and women according to total hip YAM (PR). In a report on the prevalence of osteoporosis in the lumbar spine and femoral neck in those aged ≥40 years old, estimated from the criteria of the Japanese Society for Bone and Mineral Research, the prevalence of osteoporosis of the lumbar vertebrae L2-4 is 3.4% in men and 19.2% in women, and that in the femoral neck is 12.4% in men and 26.5% in women. When comparing our results against these criteria, the prevalence of osteoporosis of the lumber spine was higher in men, although the prevalence of osteoporosis of the lumber spine was lower in women.

The prevalence of the femoral neck was lower in both men and women. (Fig. 1, Fig. 2, Fig. 3, Fig. 4, Fig. 5, Fig. 6)

3. Comments from radiologists

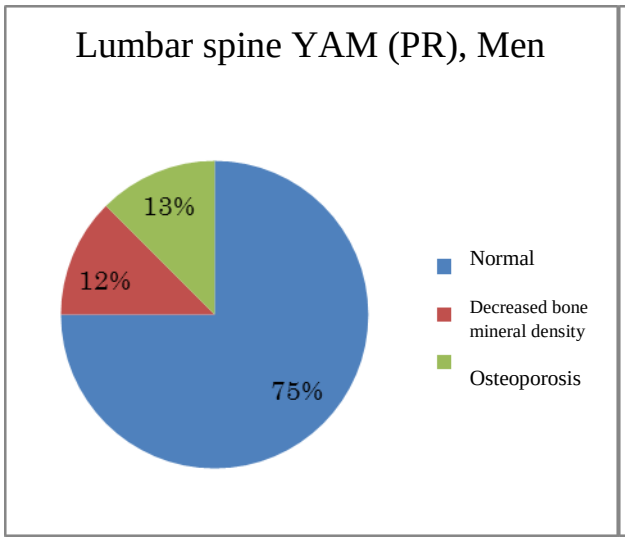


Fig. 1

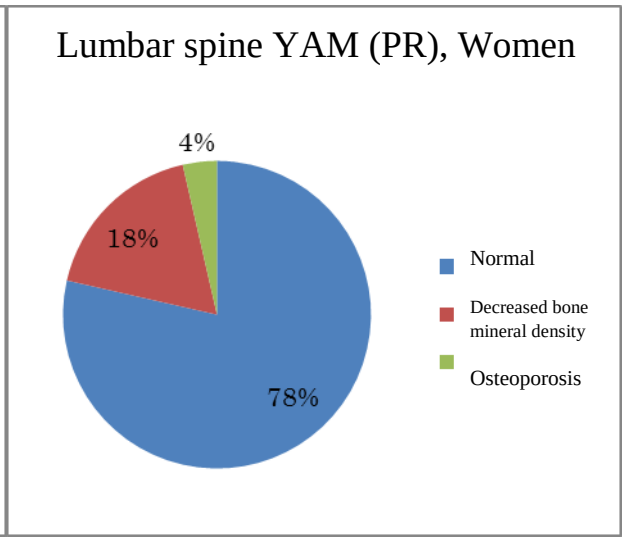


Fig. 2

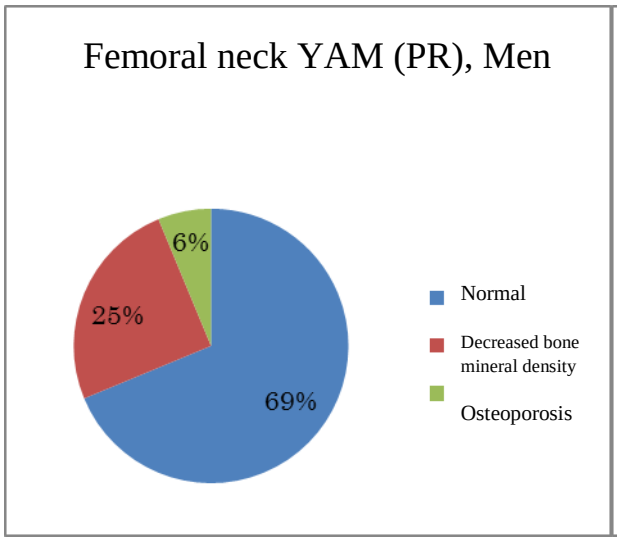


Fig. 3

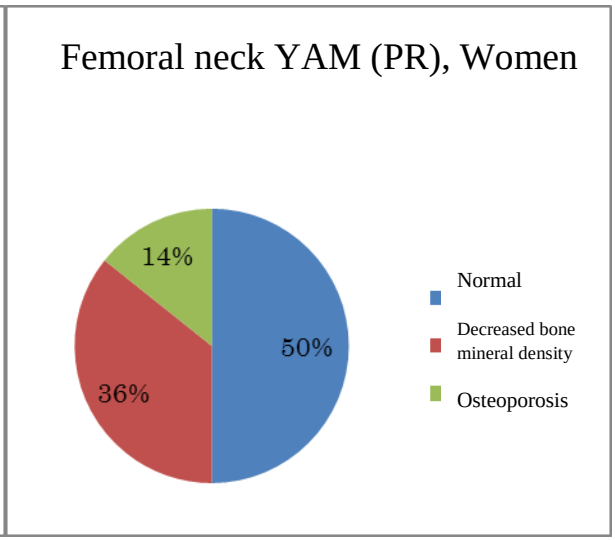


Fig. 4

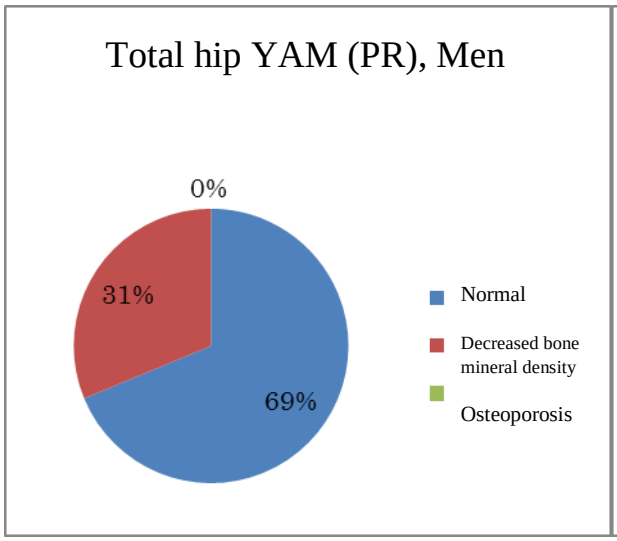


Fig. 5

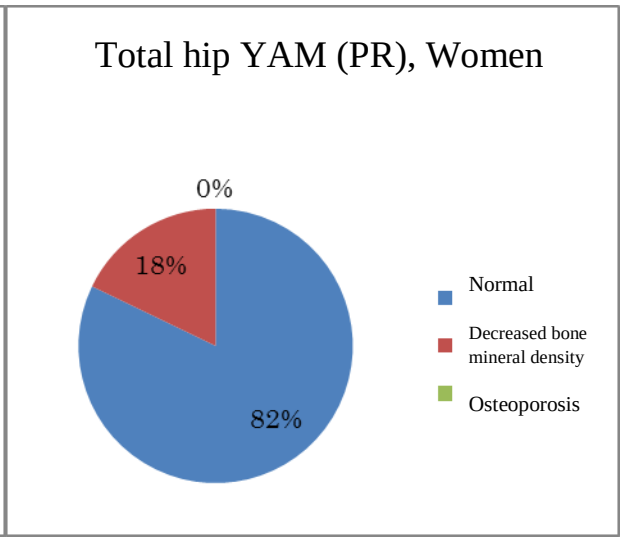


Fig. 6

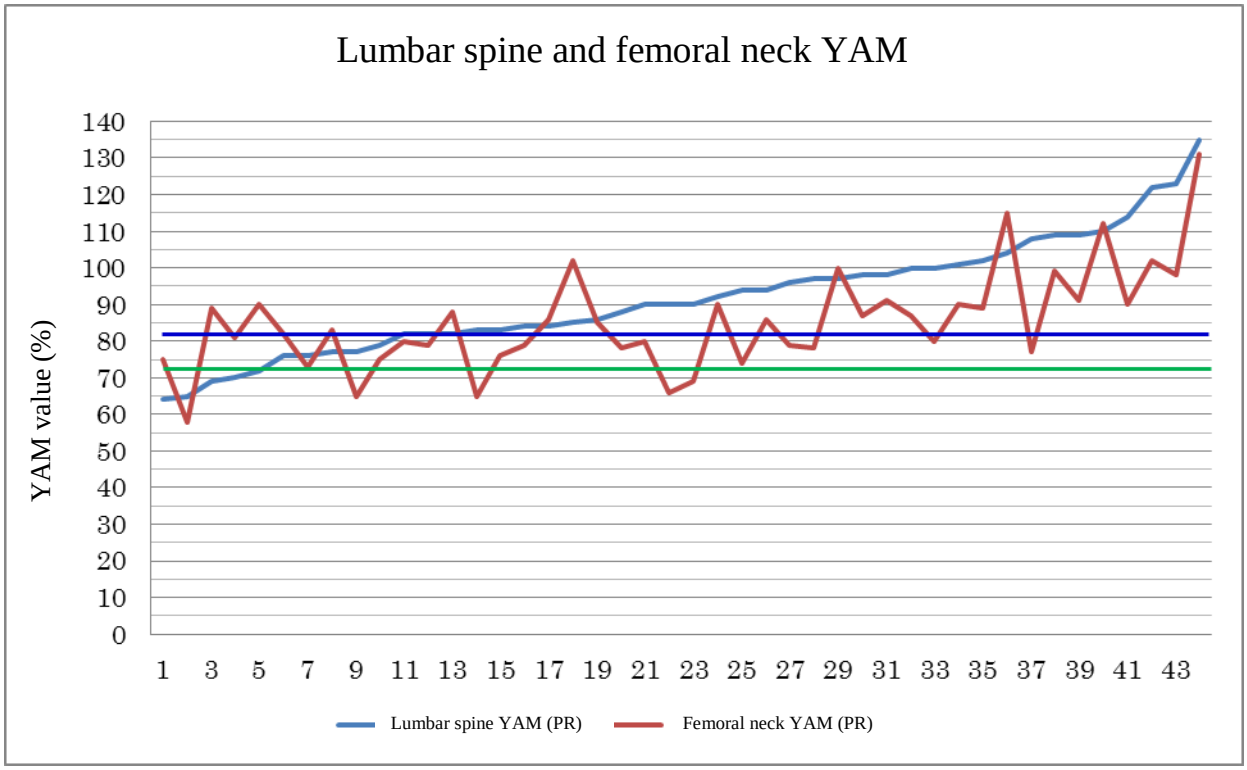


Fig. 7

3. Comments from radiologists

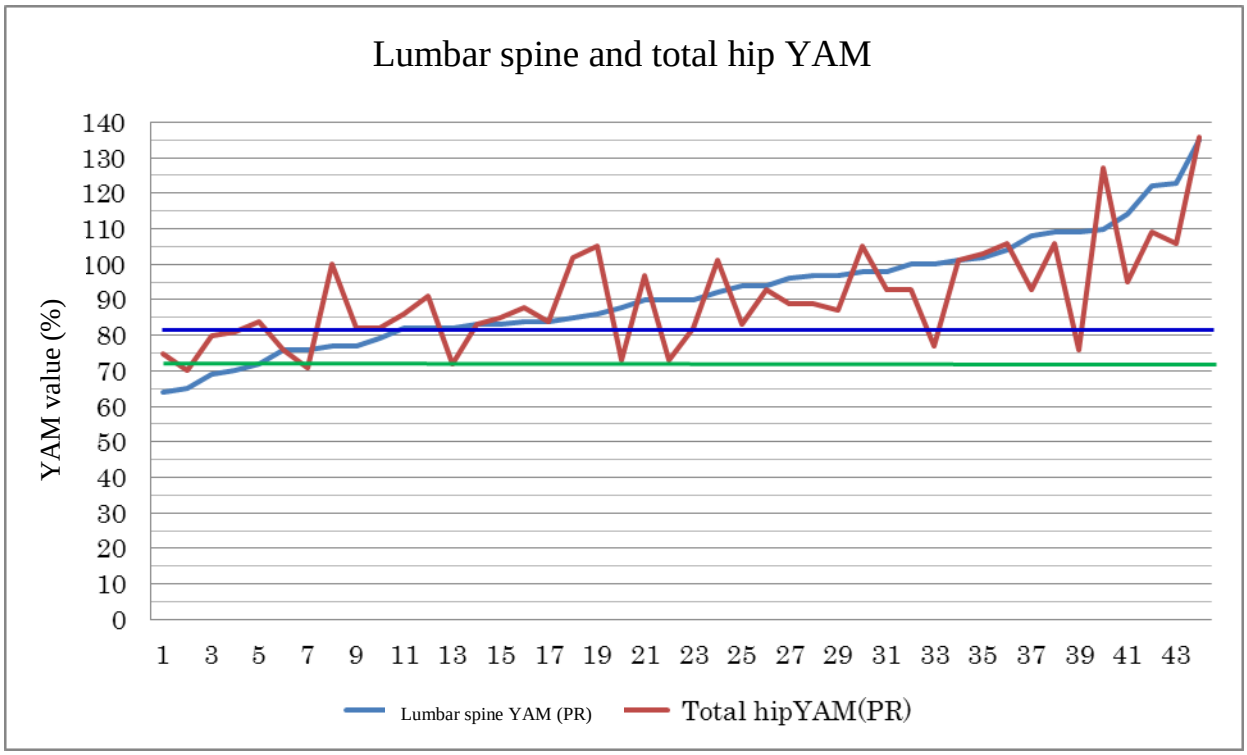


Fig. 8

YAM value	Lumbar spine and femoral neck		Lumbar spine and total hip	
	Men	Women	Men	Women
0-10	7	14	10	17
≥11	9	14	6	11

Table 2

When lumbar spine YAM is high, femoral neck and total hip YAM values also increase. However, quite a few people had YAM values of 11 or above for the lumbar spine and femur. This means there is a risk of missing abnormal values in the femur in cases where only measurement of the lumbar spine is performed. (Figs 7, 8, Table 2)

The relationship between the femoral neck and total hip is similar, with the total hip YAM tending to be higher than the femoral neck YAM. (Fig. 9)

These results suggest that both the lumbar spine and femur should be tested and abnormal values looked for when measuring bone mineral density in thalidomide-impaired patients.

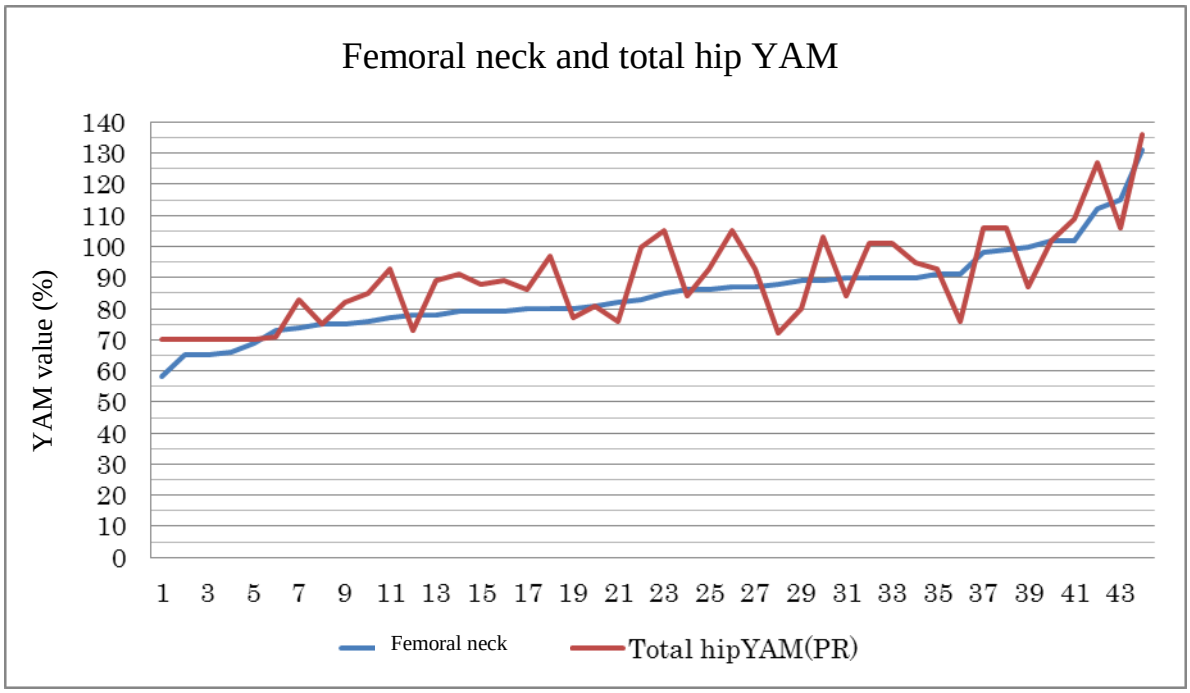


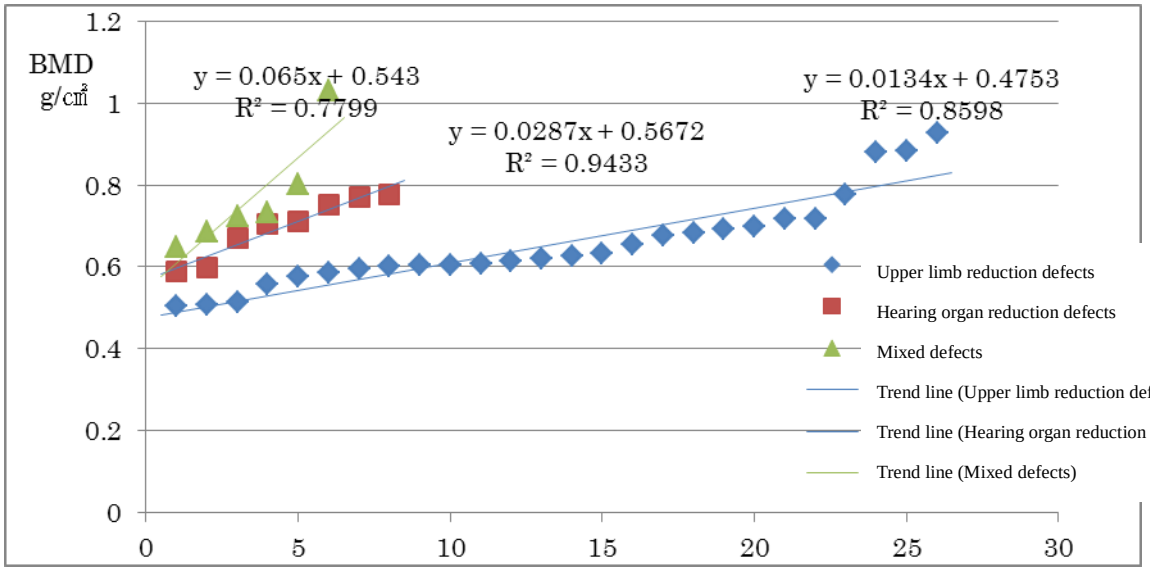
Fig. 9

Q3-4: Are there any particular trends in the bone mineral density of thalidomide-impaired patients?

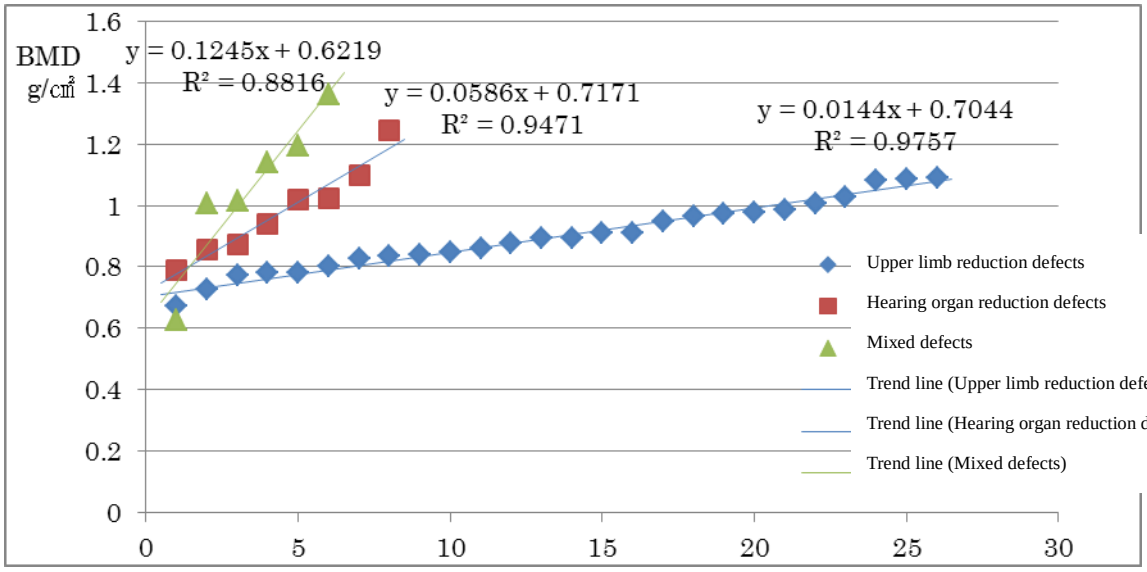
- **Bone mineral density at the femoral neck is low (YAM = 76%) in patients with upper limb reduction defects.**
- **Bone mineral density tends to be low in patients of short stature and low body weight.**

In bone density measurements in 40 thalidomide-impaired patients (upper limb reduction defects: 26 patients; hearing organ reduction defects: 8 patients; mixed type: 6 patients), low bone density was seen in about 60% of patients. When comparing the three types, bone mineral density was lowest in those with upper limb reduction defects, followed by those with hearing organ reduction defects and those with mixed defects. (Fig.1)(Fig. 2)

3. Comments from radiologists



(Fig. 1) Comparison of bone mineral density (BMD) values in the femoral neck



(Fig. 2) Comparison of bone mineral density (BMD) values in the lumbar spine

In those with upper limb reduction defects, the mean femoral neck bone mineral density value was 0.66, which is 76% when converted to the YAM value. On the other hand, the mean lumbar spine bone mineral density value was high, at 0.9. When converted to the YAM value this is 88.5%, which is unexpected in view of the changes resulting from thalidomide impairment

In infancy, which is a time of vigorous growth and development with increased bone formation and bone metabolism, patients with upper limb reduction defects would probably not have exposed the bones to sufficient loading because of restricted mobility. With insufficient bone loading, there is a paucity of calcium deposition and the marked inhibition of bone mass increases. Also, the outward

appearance of those with upper limb reduction defects would tend to lead to negative social experiences, social withdrawal and fewer opportunities to go outside. This in turn would reduce the length of time exposed to sunlight, making it difficult for the body to synthesize vitamin D that aids in the absorption of calcium, thus inviting low bone mineral density.

In terms of nutritional balance, those with upper limb reduction defects would probably have greater difficulty than normal people in attaining an adequate dietary intake, and without a balanced diet, calcium intake would decrease and bone mineral density would be unlikely to increase. When the YAM value is below 80%, the question of dietary intake must be tackled with great care and consideration.

No abnormalities of the upper or lower limbs were seen in those with only hearing organ reduction defects, and because this group was less limited in their physical activity than those with upper limb reduction defects, we would expect a tendency for their bone mineral density to be higher.

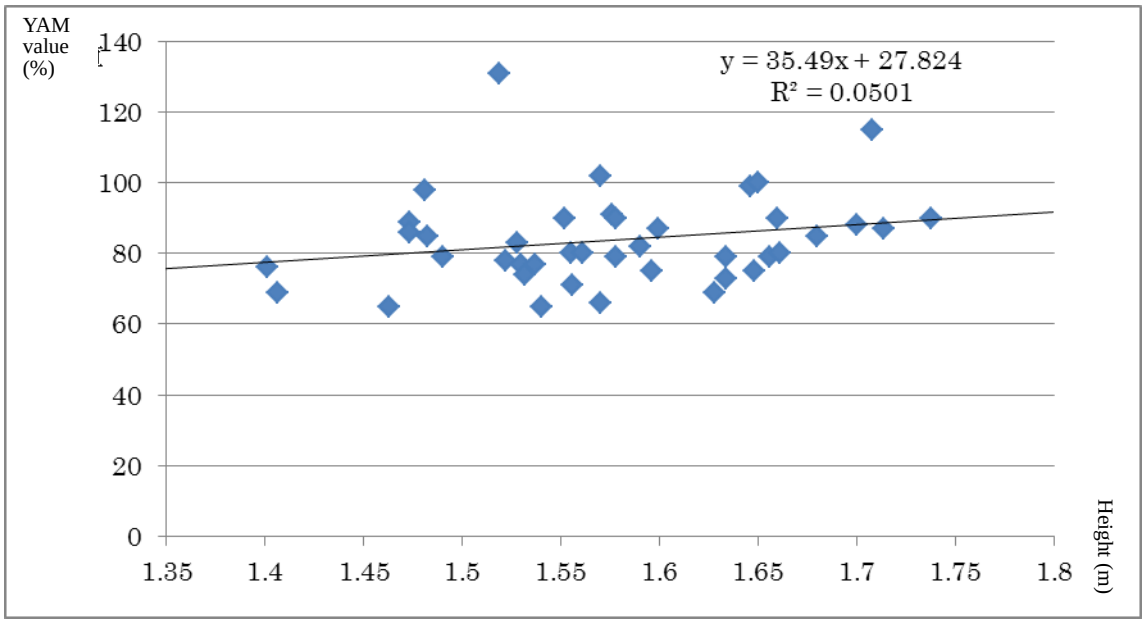
In this analysis, we found that low bone mineral density was much more pronounced in the femoral neck than the lumbar spine. Femoral neck fractures can leave patients bedbound and has a 5-year survival rate of 50%, indicating that those at risk must be carefully monitored.

Bone mineral density was lower in those of shorter stature and lower body weight. (Fig. 3, Fig. 4, Fig. 5, Fig. 6)

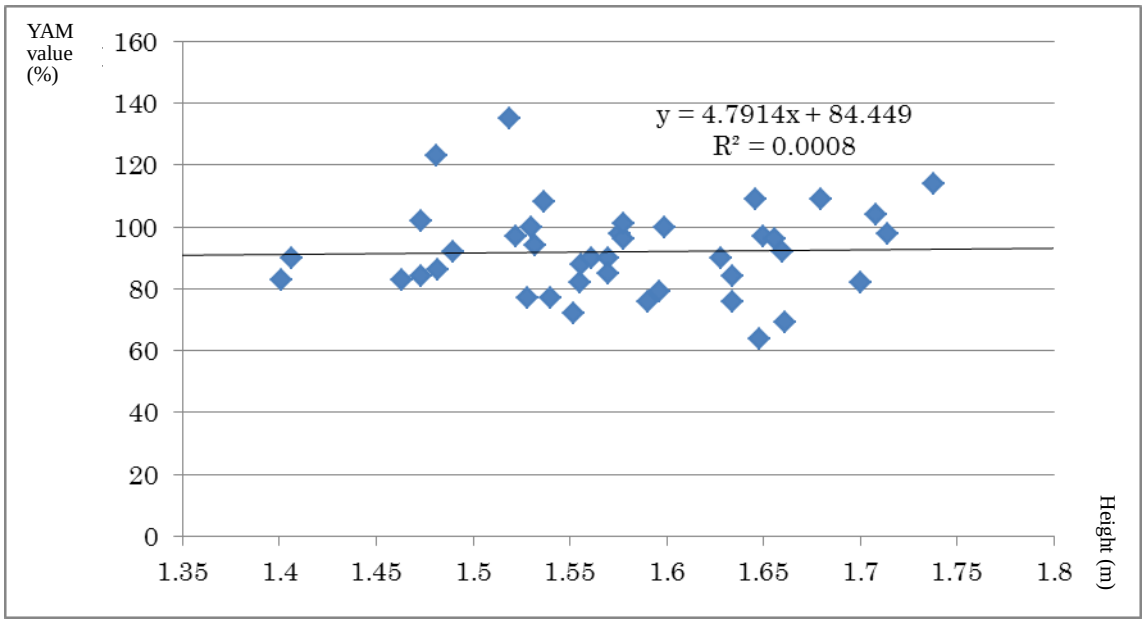
In relation to height, bone mineral density is maintained as height increases, and is low at heights of 155 cm and below. It is thought that bone mineral density is low in shorter people because of insufficient bone loading, as is the case with limited mobility.

Similarly, in relation to body weight, bone mineral density becomes higher as body weight increases because bone loading is greater. YAM values for bone mineral density in the femoral neck and lumbar spine exceeded 80% in patients above 65 kg.

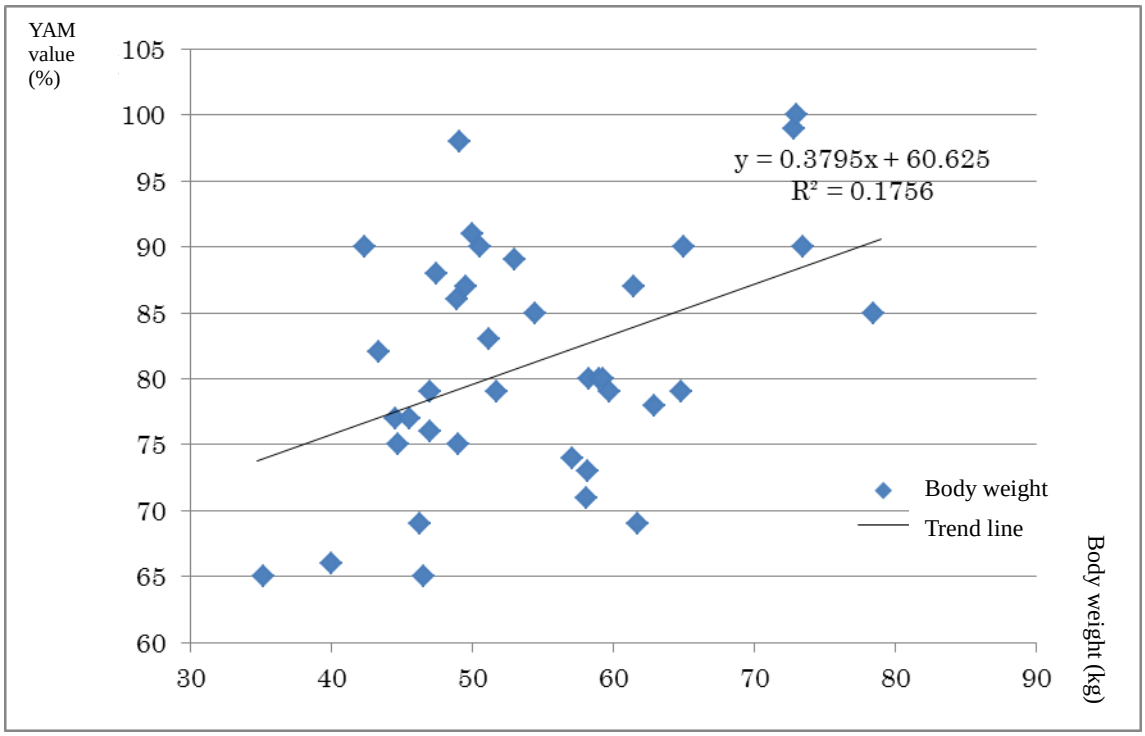
3. Comments from radiologists



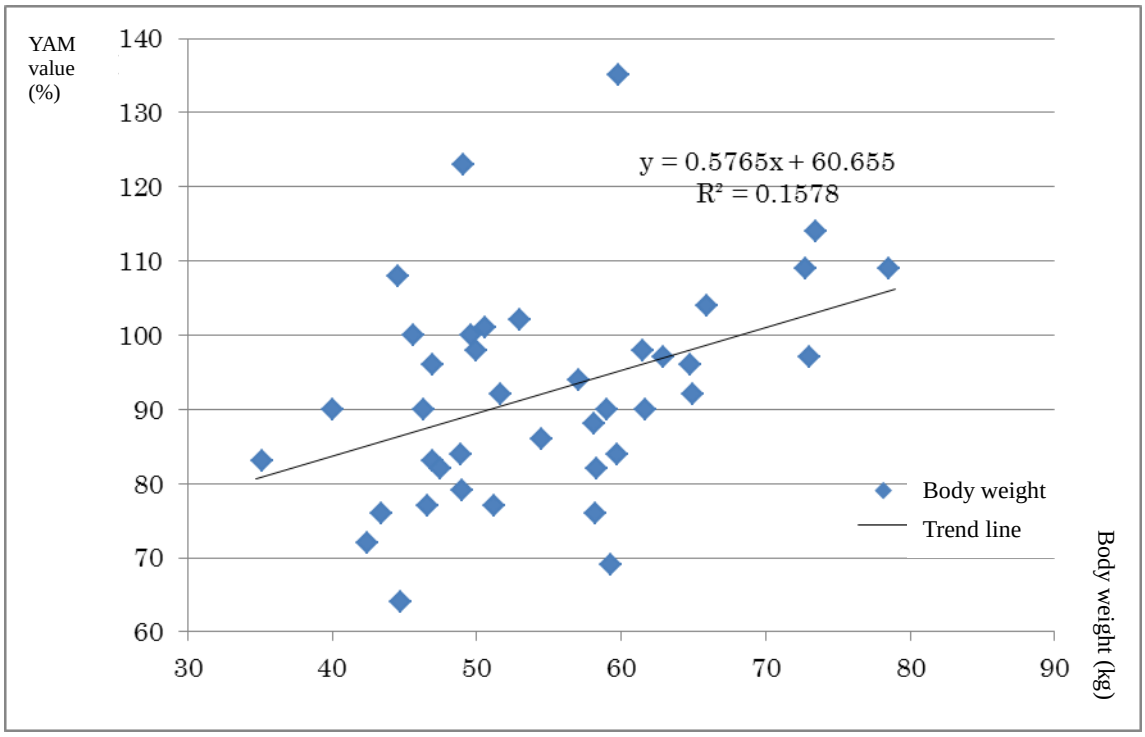
(Fig. 3) Comparison of height and YAM values in the femoral neck



(Fig. 4) Comparison of height and YAM values in the lumbar spine



(Fig. 5) Comparison of body weight and YAM values in the femoral neck



(Fig. 6) Comparison of body weight and YAM values in the lumbar spine

3. Comments from radiologists

Bone mineral density was clearly low in the femoral neck in those with upper limb reduction defects. For such patients, there is a high risk of falling, with the potential fractures having greatly detrimental effects on quality of life (QOL). We hope that by sharing these results with healthcare teams, we can help to maintain the QOL of these patients. This information also needs to be closely shared with regional healthcare centers.

As a result of this analysis, it is possible to inform patients of the high risk factors for osteoporosis, and to advise them on how to prevent osteoporosis themselves through calcium intake, physical activity, etc.

Many thalidomide-impaired patients have inevitably suffered from retarded bone development and reduced muscle strength because of the severe limitations in their activities of daily living due to functional and anatomical impairments of the upper limbs from birth. As these patients age, they will also develop secondary disabilities, but we hope that even though their ability to perform daily activities may become progressively impaired, this research will help in preventing fractures that could severely harm their quality of life.

4. Comments from occupational therapists

Main author: Takeshi Kobayashi

Q4-1: I have very stiff shoulders. Would it be alright to get a massage?

Stiffness from the neck to the shoulders is a problem for many thalidomide-impaired people, and may also include numbness in the arms, hands and fingers. Many people with shortened arms or with one arm much shorter than the other have muscle stiffness over the entire area from the neck to shoulder blades, not just in the shoulders. Massage will be effective if it can alleviate the stiffness, but you should first have your shoulders examined by, for example, the orthopedic surgery department and getting it X-rayed. Although this problem is generally called 'shoulder stiffness', it can be caused by issues such as abnormalities of the neck bones; hence, caution is needed before getting the area massaged. Even after a normal first checkup, it is a good idea to have regular tests and checkups. (See Q2-1)

Q4-2: I have pain in my lower back. Would it be alright to get a massage?

Many thalidomide-impaired people also suffer from low back pain (lumbago). Those with shortened arms, in particular, often use the whole body when reaching for things with the hands. This puts a burden on the lower back. Sufferers may also have numbness in the backs of the thighs and in the toes. Massage should be effective in loosening up the muscles, but you should first have the problem investigated by, for example, consulting the orthopedic surgery department and getting an X-ray. Although this problem is generally called 'low back pain', it can also be a result of abnormalities in the bones of the lower back, which requires caution. Even after a normal first checkup, it is a good idea to have regular tests and checkups. (See Q2-2)

Q4-3: I have developed pain in my hip joints and greater difficulty in moving. When I was examined at a hospital, I was told it was just part of the aging process, but I am worried that I may become immobile if nothing is done.

Pain and restriction of movement in the hip and knee joints are certainly a reality for many people as they get older. Women, in particular, may have hip joint impairments or one leg shorter than the other. Treatments, such as exercise therapy, may be recommended depending on the results of physical examination. In some cases, it is not only the joints that are affected, but the muscles may also have stiffened or contracted. This also requires suitable examination, and can often be

improved or prevented from getting worse by stretching. It is particularly important to prevent these problems in patients with shortened arms, as they often use their legs for performing day-to-day activities. Hence, you should have the problem investigated by, for example, consulting the orthopedic surgery department and getting an X-ray. (See Q2-5)

Q4-4: I am worried because my posture is gradually getting worse. My body is beginning to look stooped.

This is common in people with upper limb impairments, particularly those with shortened arms. Such people are prone to developing a rounded, stooping posture (kyphosis). This posture is typically caused by habitually slouching when using the hands, so that the whole of the shoulder (the shoulder girdle) sticks forwards. This can easily lead to a feeling of poor posture, fatigue in the muscles around the shoulder and neck, etc. Exercises, such as back stretching, pushing out the chest and opening the arms, are useful. These should be done slowly, with big movements, but without overstraining yourself. It is better to do a few sessions of these every day, several times a session, rather than doing many repetitions all at once. For example, you could do three sessions a day, morning, daytime and evening, performing each exercise 3 times in a session, slowly and with big movements. Try the approach that best suits your physical condition and pace. You might also get some ideas by listening to the morning exercises broadcast over the radio in Japan (*rajio taiso*).

If you feel numbness in the arms, legs or feet, or pain in the neck or lower back, please have the problem investigated by, for example, consulting the orthopedic surgery department and getting an X-ray.

Q4-5: I have recently started noticing many difficulties in my activities of daily living, such as trouble getting my arms through sleeves when getting dressed, trouble opening bottles, etc.

It is difficult to say whether these recent problems with deterioration of joint movement, loss of strength and so forth are directly related to age or to thalidomide-induced impairments. It must also be worrying to find that you are able to do less and less, especially as you have had to make great efforts to find ways of coping with everyday activities since infancy. In the future, you will probably need to continue to maintain joint mobility and muscle strength in a way that suits your own style and pace. But you should be careful not to overdo things, as you will cause yourself pain if you overburden your joints and muscles. In future, it will also be important to use equipment such as self-help aids to make things easier in your daily life while alleviating the burden on your body. If there is an occupational therapist at a local healthcare center, please ask them about self-help aids and assistive devices[Note].

Q4-6: I regularly use a computer for my job, but I am getting numbness in my hands and fingers. Recently I have had difficulty using a mouse.

The main cause of computer work-related numbness in the hands and fingers is carpal tunnel syndrome, and this is a widespread problem not restricted to thalidomide-impaired people. Nevertheless, caution is needed as thalidomide-impaired people are inherently more prone to develop carpal tunnel syndrome. Commercially available wrist pads are generally used to ensure that the wrist joints are bent downwards rather than upwards. The pad height defines the angle of the wrist joint, but everyone is different, so you should experiment with different heights and materials. The mouse can also be adapted to suit the individual, as the hands and fingers of thalidomide-impaired people typically differ in terms of length and structure. If you are able to consult with an occupational therapist with knowledge of self-help aids, they should be able to find a solution tailored to your specific needs[Note]. (See Q2-3, Q2-4)

Q4-7: I get dry eyes and have difficulty seeing around me when I move my eyes.

This is a common problem in people with impaired hearing, particularly those with paralysis of the facial muscles. These people can have various other symptoms apart from dry eyes, such as watery eyes. Firstly, you should have this investigated by the ophthalmology department. The difficulty in seeing around you when you move your eyes could be because the small muscles that

make your eyeballs move are not working enough. There are various approaches to this problem depending on the symptoms, so please arrange to be seen by a hospital or clinic that has an orthoptist. (see Q1-4).

Q4-8: I am anxious because my parents are beginning to need nursing care and I am not sure how the long-term care insurance system works.

Your parents must be around 75 years old or more, so it is understandable that they will gradually need help. In Japan, long-term care insurance is generally available to people aged 65 and over, but applicants must obtain certification by applying to the municipality where they reside. Upon application, the degree of care required is determined, which dictates the kind of service you receive. After obtaining certification, you will need to choose a care manager. The care manager will create a care plan in consultation with the person requiring care and their family. The care plan should be selected in accordance with the wishes of the person requiring care, as there are various aspects to the plan, such as day services or home visit services, provision or loan of assistive devices and aids, etc. As a general rule, copayment by the recipient for use of these services is set at 10%. Please enquire at the relevant department of your local municipality (e.g. the Senior Citizens Welfare Division).

Q4-9: Is long-term care insurance also available to thalidomide-impaired people? What other help is available?

In principle, long-term care insurance is intended for people aged 65 and over. From the age of 40, you can only receive this insurance if you have a 'specified disease' (e.g. cerebrovascular disease, Parkinson's disease, rheumatoid arthritis, etc.). Unfortunately, thalidomide impairment is not currently one of the specified diseases, so thalidomide-impaired people cannot use long-term care insurance at this age. Under the *Services and Supports for Persons with Disabilities Act*, people with disabilities could receive certification for a particular disability level classification from the municipality where they live, following a similar procedure as the long-term care insurance procedure. However, this law was replaced by the *Act for Comprehensive Support of Daily and Social Activities for Persons with Disabilities* on April 1st, 2013, so please consult with the secretary of the Ishizue Foundation for more information.

Q4-10: What is the disability pension? Is this the same as the Ishizue pension, and is it also available to thalidomide-impaired people?

The disability pension is paid to you if you became disabled by a disease first diagnosed while enrolled in a pension plan (while paying an insurance premium) or at around the age of 20. It is not the same as the Ishizue pension. There are two types of disability pension, the basic disability pension and the employees' disability pension. Both must be applied for. The documents needed for application, such as a doctor's certificate, and the center where the application needs to be made, differ for different people, with the certification criteria for the grade of disability also differing according to the disability. There are various conditions for application, so please consult with the secretary of the Ishizue Foundation before applying.

Note: Occupational therapists (OTs) are healthcare specialists who provide guidance and support, such as strategies for managing activities of daily living, for people with, or expected to develop, physical or psychological impairments. To support patients in their daily lives, OTs introduce or adapt self-help aids and assistive equipment, and devise solutions specifically tailored to the individual, as needed. You will normally need to be referred to an OT by a doctor. Please enquire at your local healthcare center or contact the secretary of the Ishizue Foundation.

5. Blood collection

Main author: Atsuto Yoshizawa

Q5-1: How do you collect blood from people whose upper limb impairments make this a difficult procedure?

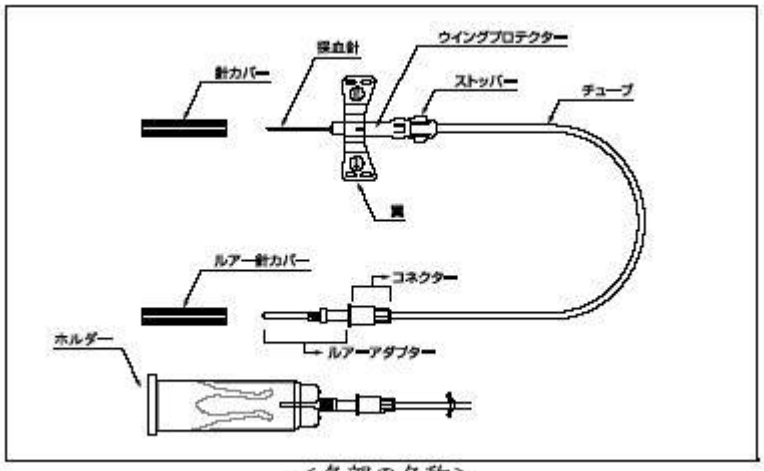
- **See attached source document 1: Techniques for taking blood from thalidomide-impaired patients**

Many thalidomide-impaired patients have had bad experiences with blood collection and are highly anxious and fearful of needles and having their blood taken. First, ask the patient if there is a site where blood collection has been unproblematic a few times in the past, and proceed to collect blood from that site. If unsure of the puncture site, check with other nurses to help determine the site. If you fail the first time, ask a doctor to do the procedure rather than making repeated puncture attempts.

If puncture difficulty is anticipated, proceed after first warming the puncture site. When taking blood from the lower limbs, it is effective to proceed after first warming the leg in a bathtub filled with hot water.

Q5-2: What sort of kit is used for blood collection?

- **We use Nipro's Safetouch PSV set with Luer Adaptor**



＜各部の名称＞

各部の名称：Names of parts　　針カバー：Needle cover

採血針：Blood collection needle　　ウィングプロテクター：Wing protector

ストッパー：Stopper　　チューブ：Tube

ルアー針カバー：Luer needle cover　　ルアーアダプター：Luer adaptor

コネクター：Connector　　ホルダー：Holder

When drawing blood using this set, air inside the tube is sucked into the first blood collection tube. Blood must therefore be collected in the second and subsequent tubes if the sampling size is small, such as for blood counts and coagulation studies.

Fig. 1 Blood can only be collected from the medial side of the first toe of the right foot in this patient (24G)

Fig. 2 Blood can only be collected from the medial side of the left knee in this patient (24G)

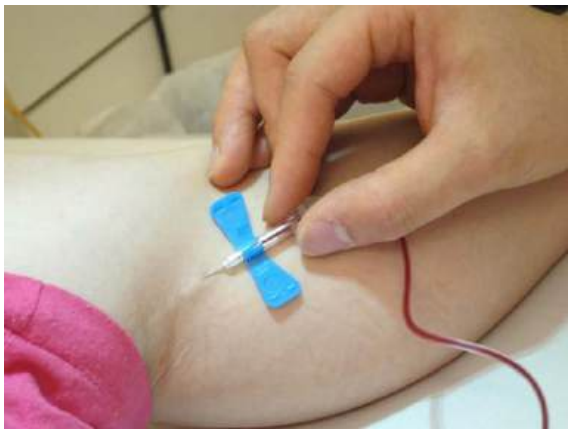


Fig. 3 Blood collection was possible from a cutaneous vein in the right upper limb in this patient (22G)

Figs. 1 and 2

Gloves are always worn as a standard precaution, but for these photographs, gloves were removed to show how to hold the equipment and the direction of the fingertip.

6. Measuring and evaluating blood pressure

Authors

Q6-1: Atsuto Yoshizawa, Hiroyuki Nagase, Yutaka Seki, Eriko Kanehisa, Takuro Shinbo

Q6-2: Yuka Shiga, Yasuhiro Maehara

Q6-3: Eriko Kanehisa, Yutaka Seki, Takuro Shinbo

Q6-4, Q6-5, Q6-6: Atsuto Yoshizawa

Q6-1: How is blood pressure measured in people with upper limb impairments?

Blood pressure (BP) is measured using an electronic BP monitor, with a cuff wrapped around the ankle so that the circle mark on the outer sleeve is lined up with the posterior tibial artery located posterior to the medial malleolus of the ankle.

We recommend taking BP at the posterior tibial artery of the lower limb using an electronic BP monitor (oscillometric method). Care should be taken because BP cannot be correctly measured unless the circle mark on the outer sleeve is aligned with the posterior tibial artery, as shown in Fig. 1. The posterior tibial artery is located posterior to the medial malleolus (Fig. 2). The pulse can be felt here by careful palpation in this area.

See Q12-2, Q12-3.

Fig. 1

Fig. 2

Q6-2: How accurate is BP measurement obtained at the posterior tibial artery using an electronic BP monitor?

- **From a study of surgical cases, we have established that BP measured at the posterior tibial artery using an S size cuff is broadly accurate.**

In 2012, we encountered a thalidomide-impaired patient without upper limbs who underwent general anesthesia together with epidural anesthesia. After entering the operating room, BP was measured every 5 minutes during surgery using a home BP measuring cuff (Terumo Corporation, S size 13 cm, 17-26 cm arm circumference) on the left leg (Fig. 3). A BP monitoring cuff that is part of the anesthesia apparatus (Nihon Kohden Corporation, 10 cm for children, arm circumference 15-23 cm) was attached to the right ankle, while arterial pressure was simultaneously invasively monitored using the dorsal artery of the right foot (Fig. 4).

For systolic BP, invasively measured arterial pressure tended to be about 10 to 20 mmHg lower than non-invasively measured BP, although both measurements showed almost identical fluctuations. The difference between BP using the oscillometric method and that by invasive measurements is believed to be about 5 mmHg, with systolic BP being lower and diastolic BP being higher in oscillometric measurements. BP in the upper and lower limbs is also said to be almost identical when measured by arterial line insertion. The differences seen in our patient were attributed to the differences between the invasive and non-invasive measurement methods.

From these results, we concluded that BP measured with an S size cuff that is suitable for the ankle circumference is broadly accurate.

Fig. 3

Fig. 4

Q6-3: Assuming there are differences between lower limb and upper limb BP using indirect measurement methods, is there a formula for estimating upper limb BP from BP measured in the posterior tibial artery?

- **When lower limb BP has been measured using an M size cuff, upper limb BP can be estimated as 0.88 × (lower limb BP + 8).**

We investigated whether upper limb BP can be estimated from lower limb BP in normal people. The ankle-brachial index is 0.9 to 1.3 in normal people, but when searching MEDLINE, we could not find a regression formula for estimating upper limb BP from lower limb BP. We, therefore, derived a regression formula for estimating upper limb BP from lower limb BP, using data from 1892 people in the 1999-2000 National Health and Nutrition Examination Survey (NHANES), an American survey that publishes analytical data, including upper and lower limb BP measurements. As a result, we obtained the formula 'upper limb systolic BP = 0.88 × lower limb systolic BP'.

We then investigated the validity of this estimation formula in 17 thalidomide-impaired patients who had been examined and for whom upper and lower limb data were available. As can be seen in Fig. 5, the estimation formula fit the data comparatively well, and we currently recommend using this formula to evaluate upper limb BP. We, however, discovered that systolic BP was being underestimated by 8 mmHg when using an M size cuff on the lower limb compared to an S size. Hence, when measuring BP in the lower limb using an M size cuff, the original systolic BP in the upper limbs is calculated by the formula:

0.88 × (lower limb BP + 8).

Caution is needed in patients with pronounced hardening of the arteries and peripheral arterial disease (PAD), because measured BP decreases and is difficult to evaluate accurately in these patients.

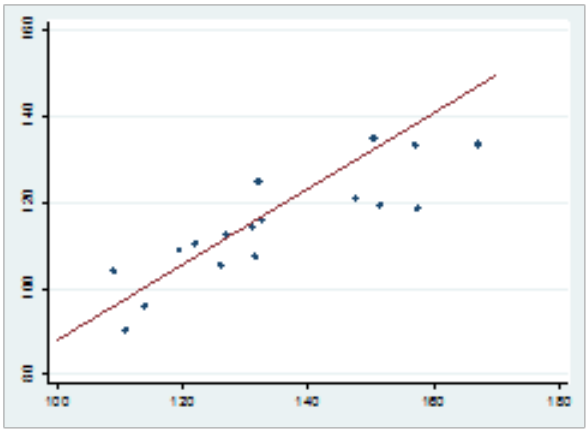


Fig. 5 Relationship between upper and lower limb systolic BP in thalidomide-impaired patients
The horizontal axis shows lower limb BP, the vertical axis shows upper limb BP, and the red line shows the estimation formula 'upper limb systolic BP = 0.88 × lower limb systolic BP'.

Q6-4: Can BP be measured in the upper limb in people with upper limbs that are underdeveloped but not completely missing?

- **If the upper arm circumference is less than 24 cm, it is preferable to measure BP using a child's cuff or S size cuff.**

The BP monitoring cuff must be of a size that fits the circumference of the patient's limb. Many people with upper limb reduction defects have had their BP measured at the upper limb, but the standard size (M size) BP monitor is intended for people with an upper arm circumference of 24 to 32 cm. It is, therefore, possible that BP measured using an M size BP monitor in patients with an upper arm circumference less than 24 cm will be underestimated. If the upper arm circumference is 17 to 26 cm, it is preferable to measure BP using a child's cuff or S size cuff to suit this circumference.

Q6-5: How should BP be evaluated in people suspected of having peripheral artery disease (PAD)?

- **If a difference is suspected between BP in the left and right lower limbs, the systolic BP in the dorsal artery of the foot and the posterior tibial artery should be measured using a Doppler blood flow meter, with an upper arm cuff wrapped around the ankle.**

If PAD is present, it is difficult to estimate systemic BP from the measured value in the stenotic artery of the lower limb. It is also possible that the arteries of both the lower limbs could be affected by stenosis. This makes it difficult to measure BP. It is, therefore, very important for

thalidomide-impaired patients to prevent hardening of the arteries.

In patients with diabetes, cardiovascular disease, etc., BP would ideally be measured in all four limbs, but in people without upper limbs, BP can only be measured in the lower limbs, and ankle-brachial pressure index (ABI) cannot be measured in this group. It should, therefore, be confirmed that there are no differences in BP between the left and right lower limbs. If a difference is detected, the femoral and popliteal arteries should be palpated to confirm whether the pulse is stronger in the left or right lower limb.

If a difference is suspected between BP in the left and right lower limbs, the systolic BP of the dorsal artery of the foot and the posterior tibial artery should be measured using a Doppler blood flow meter, with an upper arm cuff wrapped around the ankle. The higher value should be taken as the lower limb BP, and differences between left and right sides should be checked. If a Doppler blood flow meter is unavailable, it may be possible to measure systolic BP by using a stethoscope to listen to the pressure at the posterior tibial artery or dorsal artery of the foot. However, BP cannot always be assessed by auscultation in cases of PAD, and PAD, therefore, cannot be ruled out using this method.

Q6-6: Are there any home BP measurement methods that can be used without family assistance?

- **BP can be measured by wrapping the cuff around the limb while seated, then lying on your back and pressing the measurement button with your toe.**

The procedure for measuring BP at home without assistance is as follows.

1. Sit down and lightly wrap the cuff around the top of the ankle joint
2. Position the ‘○’ mark on the outer sleeve so that it is in line with the posterior tibial artery
3. Lie on your back and remain resting for 2 to 3 minutes
4. Raise your head only and push the start button using the big toe of the other foot.
5. Check the results

The equipment needs to have buttons large enough to be easily operated by the toes, and should have a cuff that can be easily wrapped around the limb using only the feet. Some patients have reported that they find the Smart Mini upper arm UA-621 BP monitor made by A&D Co., Ltd. easy to use.

7. Comments from nurses

Main authors: 16F Ward Nurses, National Center for Global Health and Medicine

Q7-1: How is blood pressure measured?

- **See Q6-1 and Q6-4.**

If it is too difficult to measure blood pressure in the upper limb, it is measured in the lower limb. The results need to be interpreted carefully, as blood pressure in the lower limb is higher than the corresponding pressure in the upper limb.

Q7-2: How are blood samples obtained?

- **See source document 1: Techniques for collecting blood from thalidomide-impaired patients**

We ask the patient which site has been most successful for collecting blood in the past, and take blood from there.

Many thalidomide-impaired patients have had bad experiences with blood collection in the past and are anxious and fearful of needles and having their blood taken. If you are unsure of the puncture site, it is best to check with other nurses before deciding on the site. If there are still problems, you should ask a doctor to do the procedure rather than making repeated attempts at puncture. If puncture is difficult, we normally warm the puncture site before proceeding. A footbath is effective for this when taking blood from the lower limb. If the patient has difficulty extending their elbow, the nurse will also assist with keeping the arm in place during blood collection. The nurse must also check that bleeding has stopped after collecting blood.

Q7-3: Are there any particular techniques for urine sample collection?

We ask how urine samples have been collected in the past, and allow patients to do this on their own if they are able to. For people who normally provide samples with a collection cup placed directly on the floor, it is fine to get them to use a collector such as a Uri-pan, if available. The patient may also need assistance in transferring urine from a cup to a centrifuge tube, pulling their trousers up and down, etc. It is a good idea to ask the patient to wear clothes that are easy to take off and put on.

Q7-4: Are there any particular techniques for abdominal ultrasound scans?

The patient may need help wiping the gel off after the scan. The nurse will also hold up the upper limbs of patients who find this difficult to do on their own.

Q7-5: Are there any particular techniques for ECG tests?

Tape electrodes may be used for the upper limbs instead of grip electrodes, depending on the extent of impairment. The test is done with the upper limb electrodes taped to the shoulders.

Q7-6: Are there any important points for hearing tests and examinations by the otolaryngology department?

Some patients with upper limb impairments have difficulty removing earwax and may therefore have blocked ears. For such patients, earwax should be removed before doing tests and examinations.

Q7-7: Are there any particular techniques for upper gastrointestinal endoscopy?

- **For hearing-impaired patients, please use source documents 2 and 3, 'Hearing-impaired patients: Upper gastrointestinal endoscopy support document'.**

It is important to inform the endoscopy room nurse of any disabilities, such as hearing loss, before the test.

Some patients with upper limb impairments tend to put themselves under pressure and feel as though they cannot breathe. You may need to give them psychological support during the test, such as by rubbing their shoulders and back. The nurse should support the patient's body if they having difficulty maintaining the lateral decubitus position and end up slipping onto their back.

For hearing-impaired patients, please use source document 2, 'Hearing-impaired patients: Upper gastrointestinal endoscopy support document'.

Q7-8: Are there any other important points relating to tests?

- **One-piece examination clothing is easier to put on and take off**

Some patients come wearing clothing that they can easily change into and out of, but others may need help changing their clothes and undergarments. If the patient needs to wear examination clothing, the one-piece gown is easier to change into and out of than the pajama-type two-piece set.

Some tests require the patient to hold the same position for a long time. If this is painful, you will need to alleviate the pain by placing a pillow, cushion or bath towel under their shoulders, feet, etc.

Q7-9: Are there any important points common to all tests?

- **It is a good idea to show the patient explanatory cards describing the test procedure before the test.**

The patient will feel less anxious and the test will probably go more smoothly if the patient is given a polite explanation of the test before starting. A communication method that the patient can easily understand should be used for the explanation, such as sign language, writing, lip-reading, etc.

The person in charge of the test should be given a description of the patient's disabilities before the test, and should conduct the test while using a mixture of written communication and lip-reading. If the test involves a number of steps, it is a good idea to show the patient explanatory cards describing the test procedure before doing the test.

See source documents 2 to 6.

Q7-10: Are there any particular techniques for medical examinations and nutritional guidance?

Examinations should be done using communication methods that the patient can easily understand, such as writing, lip-reading, gestures, etc. It is a good idea to use printed material, such as pamphlets, for giving nutritional guidance.

8. Upper GI endoscopy (via the mouth)

Main author: Toshiyuki Sakurai

Q8-1: Is the test procedure the same as for other patients?

The procedures for entry into the test room, monitoring, pharyngeal anesthesia, sedation, endoscopy, etc., are the same as usual.

Q8-2: How do you monitor blood pressure in patients with missing or underdeveloped upper limbs?

Blood pressure is measured at the posterior tibial artery in the lower limb.

Q8-3: Are sedatives necessary?

In general, it is better to use sedatives, because many of these patients will be undergoing an upper GI endoscopy for the first time and will feel greater resistance and anxiety than patients who are used to the test. Sedation is not always necessary for patients who have previously undergone this procedure. Before the test, the patient should be asked if they have previously had the test, and whether they would like sedation.

Q8-4: Are there any important points when administering sedatives?

There are two points that need to be taken into consideration. Firstly, intravenous injection in the upper limb is not possible in some patients with missing or underdeveloped upper limbs. In such cases, intravenous injection via lower limb veins is an option, but establishing a suitable vein is difficult in some patients. Many patients who have experienced repeated failed attempts at intravenous injections are fearful of the procedure, so it is important to proceed with a gentle tone of voice.

The second point relates to the injection dose. Caution is necessary in patients with missing upper limbs, as there is a possibility of using too much sedative relative to the patient.

Q8-5: What sort of endoscope is used?

In principle, any type of endoscope can be used. However, we have inserted a transnasal endoscope through the mouth in some patients who were too fearful to accept a normal diameter endoscope through the mouth. Many patients have a small physique, and we therefore select the endoscope after careful consideration of the patient's body size and level of fear.

Specifically, we have used the Olympus H260, Q260, Q240X and XQ240. We have made frequent use of endoscopes with a slightly finer diameter than normal.

Q8-6: Do patients have difficulty adopting a suitable position for endoscopy?

Assuming the left lateral decubitus position is possible even for those without upper limbs. However, tests lasting for a long time can be more painful for these patients, as this posture pushes the shoulder inwards.

Q8-7: Are there any important points when inserting the endoscope?

Thalidomide-impaired patients may have abnormalities of the pharynx, larynx and esophageal orifice , but these do not prevent endoscope insertion. A small-diameter endoscope should be used if the patient is particularly fearful of scope insertion.

Q8-8: Are there any important points regarding endoscopic observation?

There is nothing specifically requiring attention. As always, it is important to look carefully for any anatomical abnormalities, but we have not discovered anything in tests to date.

Q8-9: Are there any techniques for getting the patient to relax?

Many patients feel stressed and anxious, so do not explain things to them too quickly or in an overbearing manner. Try not to behave in a way that makes the patient feel stressed.

Q8-10: What strategies are used for those with hearing loss?

We prepare cards beforehand with a description of the test or specific instructions, such as 'We are going to start the test', 'Don't swallow any saliva', 'Breathe out', etc., and proceed with the test while showing these to the patient. (See attached documents)

Q8-11: Have there been any abnormal anatomical findings?

We have seen no noteworthy anatomical abnormalities in numerous observations from the pharynx to the duodenum.

Q8-12: Are there any other important points?

We have noticed that thalidomide-impaired people tend to feel very stressed and anxious about the tests. The person performing the test should, therefore, ideally be a highly experienced, senior physician.

It is important that the patient is relaxed for the test. It is also important to consider changing to a smaller-diameter endoscope at an early stage to suit the patient, rather than trying to force things.

Q8-13: Are there any important points for nurses attending the test?

As with the physician performing the test, nurses should try to create a relaxed and welcoming atmosphere. It can be useful to rub the patient's back, or actively communicate with non-sedated patients or show them cards with written instructions during the test.

9: Upper GI endoscopy (via the nose)

Main author: Takama Maekawa

Endoscopy was performed in 9 of 10 thalidomide-impaired patients who came for checkup in FY2012 and all 8 patients who came for checkup in FY2013− a total of 17 patients. After we explained the difference between transnasal and transoral endoscopy, 11 patients chose transnasal endoscopy and 6 chose the transoral route. Based on this experience, we prepared this Q&A for physicians and nurses performing checkups on thalidomide-impaired patients using transnasal endoscopy for the first time.

Q9-1: Is the choice between transnasal and transoral routes made in the usual way?

Transnasal endoscopy has recently been developed to provide an expanded visual field and improved resolution, and is now very close to transoral endoscopy in terms of performance. Transnasal endoscopy produces very little pain and does not require the use of sedatives. Therefore, as long as transnasal endoscopy is not contraindicated, we consider it to be the best choice for examinations, although the transoral route should, of course, be selected if the patient requests it. Two-thirds of the thalidomide-impaired patients endoscopically examined by us chose the transnasal and one-third chose the transoral route.

Q9-2: Are any patients contraindicated for transnasal endoscopy?

The nasal route is contraindicated in patients with bilateral obstructive disease of the nasal cavities and the epipharynx. Use of the nasal route may not be possible in patients with otolaryngologic diseases or after surgery for such diseases. This route can be used in patients taking antithrombotic agents, but is contraindicated if there is a tendency to bleed because of an underlying disease that involves significant reduction in platelets, such as liver cirrhosis. A history of aspirin-induced asthma is also a contraindication for nasal endoscopy, because of the possibility of induction of aspirin-related asthma due to the parabens that is used as a preservative in Xylocaine products (other than 8% Xylocaine spray), i.e. Xylocaine jelly, Xylocaine viscous and 4% Xylocaine liquid. Naphazoline nitrate nasal drops are contraindicated in patients using MAO inhibitors, because they can cause a rapid increase in blood pressure. None of the patients examined by us were contraindicated for transnasal endoscopy.

Q9-3: Is the transnasal endoscopy procedure the same as for normal patients?

It is basically the same, but we use explanatory cards to help hearing loss patients understand the test procedure.

Q9-4: Does transnasal endoscopy take the same amount of time to perform as transoral endoscopy?

For both the transnasal and transoral routes, the examination starts about 30 minutes after the patient takes Gascon and Pronase to clear gastric mucus and bubbles, in order to facilitate observation of the stomach. During this period, the transnasal route requires a little time to open up the nasal cavity and administer an anesthetic agent and a vasoconstrictor to prevent bleeding. As with other cases, transnasal endoscopy takes about 1.5 to 2 times longer than endoscopy by the transoral route from scope insertion to removal, although the longer time is not a problem because the transnasal route involves very little pain.

Q9-5: Are there any precautions to be observed when performing transnasal as opposed to transoral endoscopy?

The usual precautions apply. The endoscopist should encounter no technical problems performing the procedure as long as he/she has a thorough grounding in transoral endoscopy, and has reviewed his/her understanding of the properties of the instruments and anatomical or pathological knowledge covering the field of otolaryngology. However, compared to transoral endoscopy, the endoscopist needs to make a greater effort and adopt a more proactive attitude towards ensuring nothing is missed. The endoscopist must also adjust structural enhancement and color to make it easy to see microscopic lesions, mucosal atrophy, collecting venules, etc.

Q9-6: What brand of transnasal endoscopes do you use?

Fujifilm, Olympus and Pentax currently have transnasal endoscopes in the market. We use the Olympus GIF X-P260NX. Fujifilm has always been the leader in terms of image quality, but there is little difference between each company's latest models. It is wise to choose the latest models, as these have a 140-degree field of view, improved clarity and resolution, better water delivery and suction function and improved biopsy capture.

Q9-7: Are sedatives necessary for transnasal endoscopy?

Since transnasal endoscopy is far less painful than transoral endoscopy, sedation is completely unnecessary. All our recent patients were able to undergo problem-free examination comfortably without the use of sedatives.

Q9-8: Is pretreatment for transnasal endoscopy in thalidomide-impaired patients the same as for other patients?

The standard pretreatment for performing transnasal endoscopy requires no changes, other than

the use of explanation cards during pretreatment for patients with hearing loss.

Our center's pretreatment procedure is as follows:

1. 30 minutes before examination: The patient is given Gascon drops 5 ml + water 100−200 ml + Pronase 20,000 units + sodium bicarbonate 1g.
2. 15 minutes before examination: 0.15 ml of vasoconstrictor (Privina) is injected into the nasal cavity in order to open up and reduce swelling in the nasal cavity.
3. Nasal cavity anesthesia: We use the one-stick method. After selection of the nasal cavity with the best airflow, Xylocaine Jelly is injected into the nostril in two 2 ml doses (total lidocaine dose 80 mg). Next, a small amount of Xylocaine jelly is painted onto a 16Fr transnasal endoscopy pretreatment stick, which is then inserted 8−9 cm into the nasal cavity with best airflow and removed after 90 seconds. Pharyngeal anesthesia just before examination is unnecessary.
4. Administration of antispasmodic agent: Buscopan is not essential, but if used, can improve the quality of examination.

Q9-9: How do you choose between left and right nasal cavities?

The standard methods can be used. The side with the best airflow is chosen by inhaling through each nostril, with the opposite nostril pressed in turn. If there are any difficulties, airflow can be objectively evaluated using the nasal breathing CD disc method. We encountered no patients in who we needed to change to the opposite nasal cavity or from the transnasal to transoral route.

Q9-10: Are there any problems with body position during transnasal endoscopy?

The same position is used as for transoral endoscopy with no problems.

Q9-11: Were there any anatomical abnormalities or characteristic abnormal findings?

Different otolaryngologic regions can be observed via each route. Transnasally, the nasal cavity and epipharynx on the left side can be examined, but part of the oral cavity and mesopharynx that are visualized transorally cannot be observed. We saw no anatomical abnormalities in these examinations. The epipharynx had no notable abnormalities. In the stomach, 9 of 17 patients had no atrophy and no history of H.Pylori infection, and 4 of these had GERD L-A grade A. Four patients had closed-type atrophic gastritis and 4 had open-type atrophic gastritis, but neither of these were characteristic findings.

Q9-12: Did any patients have nosebleeds?

None of these patients had nosebleeds, and, in most cases, nosebleed can be stopped by the application of pressure. If bleeding continues, standard hemostatic methods can be used, such as

nasal sponges.

Q9-13: Do these patients need any particular kind of assistance?

It is important to help patients relax by treating them in a friendly manner. Further, hearing loss patients should be given an easy-to-follow explanation using explanation cards. Sedation is completely unnecessary, but physical touch, such as rubbing the patients back, for example, is effective in getting them to relax. Since the transnasal route is far less painful than the transoral route, patients might also find the examination more relaxing if they are given an explanation of the procedure while watching it on a submonitor, if they so desire.

10. Anesthesia

Main authors: Yuka Shiga, Yasuhiro Maehara

Q10-1: Are there any particular points to be aware of in preoperative rounds?

The usual matters should be noted: previous history, history of present condition, physical findings, test findings, etc. However, for physical findings in particular, lower limb IV sites and monitor positioning should be confirmed in advance if the patient has underdeveloped or missing upper limbs. Insertion of a central venous line may need to be considered, because securing vascular access in the lower limbs can be difficult. This should be done while speaking to the patient in a calm voice. If epidural or spinal subarachnoid anesthesia is to be given, it should be confirmed that the patient can adopt the necessary posture.

Q10-2: Are there any contraindications for particular anesthesia methods?

There are no particular contraindications, but the method should be decided according to the patient's comorbidities and physical condition.

Q10-3: Is premedication necessary?

Patients who are undergoing surgery for the first time may feel greater stress and anxiety than normal patients. Premedication can be used in such cases, but this can cause delayed postoperative awakening. It is good to talk with the patient before surgery to find out whether they want premedication.

Q10-4: Are any particular preparations necessary?

Monitors and medication should be prepared in the usual way according to the condition and comorbidities of the patient. Since many patients are of small stature, smaller tubes, needles and monitoring equipment should be prepared to suit the patient. It is also useful to have an ultrasound scanner to confirm the course of blood vessels.

Q10-5: How do you monitor blood pressure in patients with missing or underdeveloped upper limbs?

Blood pressure is monitored at the posterior tibial artery with a cuff wrapped around the lower limb. If blood pressure cannot be easily measured in the lower limb, invasive monitoring at a large artery (femoral artery, etc.) must be considered.

Q10-6: What strategies are used for those with hearing loss?

It is a good idea to explain the anesthesia procedure on paper and with diagrams in the preoperative rounds, and to show the patient cards prepared in advance with specific written instructions ('Breath deeply', 'You will soon feel sleepy', etc.) while performing anesthesia on the day of surgery.

Q10-7: Is the amount of anesthetic agent the same as for other patients?

The amount of agent administered is determined by age and body weight, but there is evidence that thalidomide-impaired patients require smaller amounts of anesthetics. The amount of anesthetic must be adjusted while properly monitoring the patient.

Q10-8: Are there any points to consider during intubation?

There don't seem to be any particular difficulties with intubation, although many of these patients have a small physique. The diameter and insertion depth of the intubation tube should be carefully decided based on photographs and physical findings.

Q10-9: Are there any important points to consider after surgery?

As with other patients, postoperative circulatory and respiratory condition should be monitored and pain management should be provided, as necessary.

11. Psychiatry

Main author: Kobun Imai

Q11-1: What kinds of psychological and psychiatric problems do thalidomide-impaired patients have?

From June to December 2012, the Ishizue Foundation for thalidomide-impaired people conducted a survey questionnaire (effective number of respondents: 201) on the current living conditions of 286 thalidomide-impaired people whose addresses were known. The survey found that 64.5% (130) of all respondents said that their physical condition was poor (subjective symptoms) because of illness, injury, etc. Among the 130 people with subjective symptoms, 16.2% (21 respondents) had visited a health center because of 'depression or other emotional disorders'.

In Japan, mental health problems in thalidomide-impaired people have been under continuous assessment since the 1990s. In a survey (Saito, 1997) by the Ishizue Welfare Organization of 296 thalidomide-impaired people (160 men, 136 women) using the GHQ-28 questionnaire, there was almost no correlation between points scored on GHQ-28 and the level of each disability (overall, limbs, hearing). Unemployed survey subjects did, however, have significantly higher overall scores on GHQ-28 than the employed, while unmarried subjects scored significantly higher than married subjects on indicators measuring the subscale 'depressive state'.

When the Mini-International Neuropsychiatric Interview (M.I.N.I) was administered to 22 thalidomide-impaired people examined at the National Center for Global Health and Medicine (NCGM) in FY2011, the results showed no correlation with specific psychiatric disorders, but 40.9% (9) of these people were suspected of psychiatric disorders of various types. We, therefore, think that as part of routine medical care, patients should, if necessary, be referred to a specialist, in addition to being given primary care for mental health issues.

Q11-2: Is there any connection between thalidomide impairment and autism?

A survey of 100 thalidomide-impaired people in Sweden found autism in 4% of subjects (Stromland K, et al. 1994). Stromland et al. considered this to be a high incidence of autism, and predicted from the clinical findings (outer ear anomalies, normal limbs) that exposure to thalidomide had occurred from the 20th to 24th days of pregnancy. On this basis, they proposed that autism starts in early pregnancy. Continuing this research, Rodier (2000) proposed the hypothesis that genes expressed at a particular period in the development of the nervous system contribute to autism. However, this contribution only appeared to apply to some types of autism.

We investigated the relationship between thalidomide impairment and autism in 22 thalidomide-impaired patients examined at the NCGM in FY2011, using the Japanese edition of the Autism-Spectrum Quotient (AQ) (Wakabayashi, 2004), but found no patients scoring 33 points or above, a score that would lead to suspicion of the presence of autism. Neither did these results show any significant difference when compared to the mean overall AQ scores and subscale items in normal subjects.

Q11-3: Is epilepsy common in thalidomide-impaired people?

Although there has been very little research using electroencephalograms (EEG) in thalidomide-impaired people, a study of 137 thalidomide-impaired people (Kanno et al, 1987) found that a history of epileptic seizures, or no history of seizures but EEG abnormalities, occurred with higher incidence in this group than in the general population, and that the incidence was high among those with hearing impairments and intellectual disabilities in particular. This result suggests that thalidomide might cause functional as well as anatomical abnormalities.

Four (18.2%) of the 22 thalidomide-impaired patients examined at the NCGM in FY2011 had EEG abnormalities, one of whom was receiving treatment for epilepsy. However, the number of patients examined in this case was too small for a comparison of abnormalities with the general population. Furthermore, in head MRI scans, all of those with EEG abnormalities had abnormal findings thought to be unrelated to thalidomide. These results suggest that the abnormal EEG findings were unlikely to be directly related to thalidomide. This survey also found no relationship between frequency of EEG abnormalities and presence or absence of hearing impairment and intellectual disabilities.

Q11-4: Are there any important points when meeting thalidomide-impaired people?

Although grouped together under a single term, thalidomide-impaired people actually have a range of levels of impairment affecting different parts of the body, and can be encountered in all walks of life. Some have difficulties with daily life and social activities, whereas others do not. It is, therefore, important to remember to respond flexibly to thalidomide-impaired individuals according to the site and extent of impairment, their life circumstances, etc.

In a survey questionnaire on the current living conditions of 286 thalidomide-impaired people from June to December 2012, the proportion of respondents with 'depression or other emotional disorder' was 5 times greater than normal for their age group, with many respondents citing health problems and anxieties about the future as the things they were distressed or troubled by in their lives. Healthcare workers need to be aware of the possibility of psychological or psychiatric problems among these patients, and should put patients in contact with an

appropriate specialist institution or agency if they see signs of such problems.

Many thalidomide-impaired people get through their daily lives with social as well as physical handicaps. It is important that healthcare workers always respect the fact that the thalidomide-impaired people they encounter not only live with their disabilities, but also overcome their disabilities in their daily lives, and that these disabilities are the result of historical medication-induced harm.

12. Other questions

Author: Atsuto Yoshizawa

Q12-1: What did you learn from the examination of 76 thalidomide-impaired patients?

- **We found that some patients had block vertebrae while others had various structural abnormalities of the middle and inner ear. We are now able to diagnose high blood pressure in patients with upper limb defects, having fixed on a method of measuring blood pressure in these patients.**

In addition to the standard tests used in the one-day medical checkups, we aimed to investigate whether these patients had any previously unknown abnormalities of the internal organs, and to determine an accurate way of measuring blood pressure in those with missing or underdeveloped upper limbs. The three key results are presented below. Some of the specialist medical terminology and medical details may be difficult for the non-specialist to understand.

1) Block vertebrae were seen in 9.3% of patients, all of whom had upper limb defects

We examined the cervical vertebrae of 76 patients and found block vertebrae in 7 of them (9.3%). All seven had upper limb defects and none had hearing impairments. The vertebrae of the cervical spine (neck bones) are normally independent of each other, but in block vertebrae, some of these vertebrae have fused together (Fig. 1). This can occur as a result of injury or tuberculosis, but is thought to be a birth defect in thalidomide-impaired patients. The presence of block vertebrae is thought to restrict the range of neck motion, weaken muscle strength in both upper limbs and cause headache and neck pain. From the age of 30 onwards, it can also cause neurological symptoms due to cervical cord compression. We wondered if block vertebrae were responsible for the long-term deterioration in body movement that some thalidomide-impaired patients complain of. We, therefore, concluded that the cervical spine should be examined in checkups for thalidomide-impaired patients, even though this is not done for other patients.

Ten (13.1%) of the 76 patients had congenital absence of the gallbladder, and 4 of these had block vertebrae.

Fig. 1

2) We found a range of structural abnormalities of the middle and inner ear

When we examined hearing-impaired thalidomide patients for structural abnormalities of the middle and inner ear by head CT, we found various abnormalities, including hypoplasia of the auditory ossicles, hypoplasia of the semicircular canals and cystic changes in the vestibules. It is possible that inner ear abnormalities may be the cause of the dizziness of which some thalidomide-impaired patients complain.

3) We decided to measure blood pressure at the ankle in patients with upper limb defects

There was no established method for measuring blood pressure in patients with undeveloped upper limbs, which meant that these patients could not know their own blood pressure. Blood pressure is one of the basic pieces of information for understanding a patient's condition, and when performing endoscopy and surgery it is essential for the doctor to know the patient's blood pressure. Therefore, we measured blood pressure in both arms and both ankles, with the goals of:

1. Determining a method of measuring blood pressure that would be able to clearly identify patients with high blood pressure
2. Determining a method that could be used by patients diagnosed with high blood pressure to monitor their own blood pressure at home.

We concluded that the best method was to measure the pressure at the posterior tibial artery at the top of the ankle joint using an automatic blood pressure monitor.

Q12-2: How high does blood pressure measured at the ankle at home have to be for it to be considered hypertension?

- **Hypertension is very likely if systolic blood pressure reaches 145 mmHg**

Blood pressure measured at the hospital tends to be higher than when measured at home. The Japanese Society of Hypertension states that in middle aged people with no diabetes or cerebrovascular disorders, blood pressure measured at home is considered to be high if it is 135/85 mmHg or above, and normal if below 125/80 mmHg. Lower limb blood pressure appears slightly higher than upper limb blood pressure, and is highly likely to be hypertension if lower limb blood pressure is 145 mmHg or above, based on systolic pressure being (135/0.88) – 8 = 145 mmHg. Patients with this level of blood pressure should discuss the possibility of hypertension with their family doctor, showing them Chapter 6, 'Approaches to blood pressure measurement'. The criteria for assessing diastolic blood pressure have not yet been established.

Q12-3: Where is the posterior tibial artery located?

- **The posterior tibial artery is located behind the medial malleolus of the ankle (Fig. 2)**

Care should be taken, because if the cuff is wrapped so that the ring on the cuff is at the lateral malleolus, as shown in Figure 4, the measurement might not display or will be inaccurate.

Fig. 2

Figure 3 Correct wrapping of cuff

==========

Figure 4 Incorrect wrapping of cuff

Q12-4: How do you deal with the fact that blood pressure measured at the lower limbs can differ between left and right?

- **Hardening of the arteries seen in peripheral arterial disease causes poor blood circulation in the legs and can cause further medical problems. Patients concerned about this should consult their family doctor.**

People with diabetes and cardiovascular diseases are at risk of peripheral arterial disease, in which hardening of the arteries reduces blood circulation in the legs, potentially causing further medical problems. Blood pressure should ideally be measured in all four limbs, but this is not possible in people with undeveloped upper limbs. You should, therefore, discuss with your family doctor if you think there may be a difference in blood pressure in your left and right lower limbs.

After getting your family doctor to show section 6-5 of this Q&A to a doctor in the cardiology or cardiovascular surgery department, it may be possible to get your blood pressure measured using a device called a Doppler blood flow meter to measure systolic pressure at the dorsal artery of the foot (at the instep) and the posterior tibial artery, with a standard arm cuff wrapped around your ankle. Usually, the lower limb blood pressure is taken to be whichever is the higher value in these two arteries. This method shows whether there are actually any differences in blood pressure between left and right lower limbs. However, Doppler blood flow meters are only used in general clinics and specialist departments. If a Doppler meter is unavailable, it may be possible to measure systolic pressure using a stethoscope to listen to the pressure in the posterior tibial artery or dorsal artery of the foot, with an arm cuff wrapped around the ankle. However, sounds are sometime audible and sometime inaudible with a stethoscope when peripheral arterial disease is present. It is, therefore, not possible to rule out peripheral arterial disease simply by measuring blood pressure in the lower limbs with a stethoscope.

Because arterial hardening occurs throughout the body, hardening can be assessed using an ultrasound device to measure the amount of thickening of the carotid artery in the neck. This device can also be used to assess other blood vessels.

Q12-5: Have similar large-scale medical examinations and surveys been done in the United Kingdom and Germany?

- **Living conditions have been surveyed, but it appears that large-scale medical examinations have not been done. The results of surveys in the UK and Germany were provided to the Ishizue Foundation after being translated into Japanese, partly funded by**

a Grant-in-Aid for Research on Regulatory Science of Pharmaceuticals and Medical Devices from the Ministry of Health, Labour and Welfare, Japan.

1) Background and survey results from United Kingdom

http://www.thalidomidetrust.org/SecureDocs/HealthGrantYear3FinalReport.pdf

2) Background and survey results from Germany

http://www.contergan.de/671/files/20130125124745Contergan_Endbericht_final_20130108.pdf

Q12-6: Have other countries also compiled a detailed Q&A on health problems in thalidomide-impaired people?

- **Possibly, but we have not been able to find any.**

We investigated this on the assumption that thalidomide victims in other countries would have had similar problems, but we found no detailed Q&A collections relating to health problems in thalidomide-impaired people. However, the UK's Thalidomide Trust has a relevant website that also covers lipid metabolism abnormalities and blood pressure measurement. We have aimed to put together a detailed Q&A with reference to this website, and we hope that by translating this Q&A into English and German it will be useful to patients in other countries as well.

http://www.thalidomidetrust.org/healthinfo

Q12-7: When I go for medical appointments, the doctors say that they do not know about thalidomide-induced disabilities and ask me if there are any useful reference materials or articles. What should I say?

Please tell them about this Q&A.

Q12-8: Do thalidomide-impaired people have any particular health problems to be aware of?

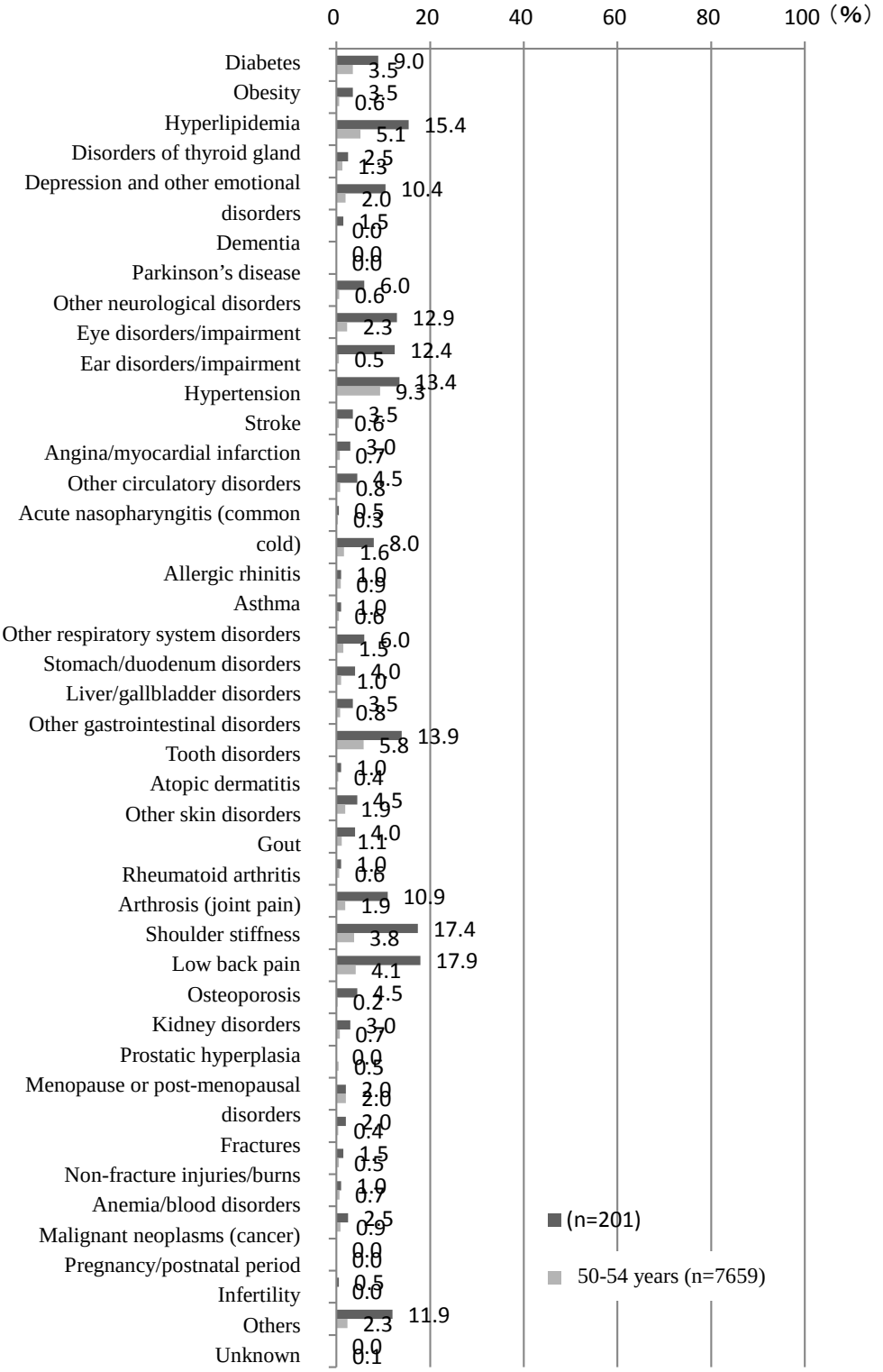
- **We strongly recommend that family doctors conduct regular checkups to determine whether patients have hyperlipidemia (dyslipidemia), hyperuricemia (high uric acid in blood) and diabetes.**
- **Our questionnaire survey found that the rates of diabetes, hyperlipidemia (dyslipidemia) and obesity in thalidomide-impaired people are 2 to 3 times greater than normal for their age group (Figure 5).**
- **Of the 76 patients who were examined, 40(52.6%) had fatty liver and 17 (22%) had hyperuricemia.**

Although there are many aspects of disability-related pain and suffering that cannot be eliminated, future illness can be prevented. The questionnaire survey results showed that many patients have diabetes, hyperlipidemia (dyslipidemia) and obesity. Fig. 5 shows the results of a FY2012 questionnaire survey on the question of the kind of health problems for which the patient regularly visited a doctor.

The most common conditions overall were low back pain (27.7%) and shoulder stiffness (26.9%). The rate of hyperlipidemia (dyslipidemia) was also very high, at 23.8%. Rates of diabetes and hyperlipidemia (dyslipidemia) were 2 to 3 times higher than normal for their age group. Of the 76 people who participated in examinations over 3 years, 40 (52.6%) were diagnosed with fatty liver and 22% had hyperuricemia (serum uric acid >7.0 mg/dl).

There may also be a considerable number of people with hidden high blood pressure, since methods for measuring blood pressure in those with upper limb defects have not yet been established. Hyperlipidemia (dyslipidemia) and diabetes are diseases that are diagnosed from blood samples. We strongly recommend that family doctors conduct regular checkups to determine whether patients have hyperlipidemia (dyslipidemia) and/or diabetes.

Fig. 5: Health problems necessitating outpatient treatment: Comparison of normal patients in their 50s and thalidomide-impaired patients (multiple responses)



Q12-9: Is the ideal body weight for people with undeveloped upper limbs equivalent to that in the general population? How do I know whether or not I am obese?

- **People with undeveloped upper limbs should not be thought of in terms of ideal body weight. It is important to measure the waist circumference as well as body weight.**

Stricter criteria should probably be applied to those without upper limbs than for the general population. This means that a body mass index (BMI) of 25 or above is a reliable indicator of obesity. However, there is no obvious answer to the question of ideal body weight in those with missing or underdeveloped upper limbs, since the extent of underdevelopment varies. Therefore, rather than asking whether a person with undeveloped upper limbs is ‘obese’, it is probably better to ask whether they have ‘metabolic syndrome’, as this can be diagnosed even if body weight is uncertain. A waist circumference of 85 cm or more for men and 90 cm or more for women is in itself a warning sign. In the recent checkups, 17 (24%) of the 66 people whose waists were measured exceeded these thresholds.

BMI is calculated as body weight (Kg) divided by height (m) squared, and in Japan obesity is defined as a BMI of 25 or above. It is recommended that patients with this level of BMI go on a diet regardless of whether or not they have upper limb defects.

It is a little known fact that the Japanese medical community distinguishes between 'obesity', ‘pathological obesity’ and ‘metabolic syndrome’. A person with a BMI over 25 is considered 'obese', but they qualify as pathologically obese if then diagnosed with central obesity (also known as abdominal obesity or visceral obesity) based on an abdominal CT scan. People with a BMI of 25 or above are also considered pathologically obese if they have one or more of the following: diabetes, dyslipidemia, high blood pressure, fatty liver or low back pain.

The FY2012 questionnaire survey found that many respondents were troubled by low back pain, so a BMI of 25 would be sufficient to qualify those people as 'pathologically obese'. In simple terms, ‘pathological obesity’ is ‘obesity’ accompanied by an illness requiring treatment.

http://www.mhlw.go.jp/topics/bukyoku/kenkou/seikatu/himan/inspection.html

In contrast, ‘metabolic syndrome’ is a set of criteria established to help predict and prevent arterial hardening. A waist circumference of 85 cm or above for men and 90 cm or above for women corresponds to 100 cm^3 or over of visceral (abdominal) fat, and if this is accompanied by any two of

high blood sugar, abnormal lipid (fat) metabolism and high blood pressure, then a diagnosis of 'metabolic syndrome' is made. This is unrelated to BMI. People with undeveloped upper limbs who have the waist circumference given above are strongly urged to get examined for abnormally high blood sugar and lipid levels, even if they have a BMI below 25.

Rather than asking whether they are obese, thalidomide-impaired patients, particularly those with undeveloped upper limbs, should therefore think in terms of whether they have metabolic syndrome, which can be diagnosed even if body weight is uncertain. This means that measuring your waist circumference is very important. You can measure your waist with the help of this site.

http://www.mhlw.go.jp/bunya/kenkou/metabo02/kiso/check/

Q12-10: Are people with undeveloped or underdeveloped upper limbs susceptible to gaining weight?

- **This is probably true, because such people have low muscle mass and, therefore, consume relatively little energy.**

It appears that many people with upper limb defects do not exercise regularly. Also, people with undeveloped upper limbs have less muscle mass than the general population, and hence, are thought to burn up less energy.

In general, energy intake (kcal) = ideal body weight × amount of physical activity

- Ideal body weight (kg) = height (m) × height (m) × 22
- Amount of physical activity:

  Light activity (e.g. desk work) 25−30 kcal × ideal body weight

  Moderate activity (e.g. standing work) 30−35 kcal × ideal body weight

If the above formula is applied to someone 155 cm tall who does desk work, then the person's energy intake should be = 1.55 × 1.55 × 22 × 30 = 1585 kcal, for which 1600 kcal is obviously sufficient. People with undeveloped upper limbs have less muscle mass and are, therefore, thought to burn up less energy than the general population. So somebody with undeveloped upper limbs who is

155 cm tall is likely to become obese if they consume 1600 kcal or more a day. Such people might well feel that they put on weight easily despite not eating very much.

Q12-11: I worry about having my blood taken because it is always difficult. Are there any leaflets or information I can show the nurses when they take my blood?

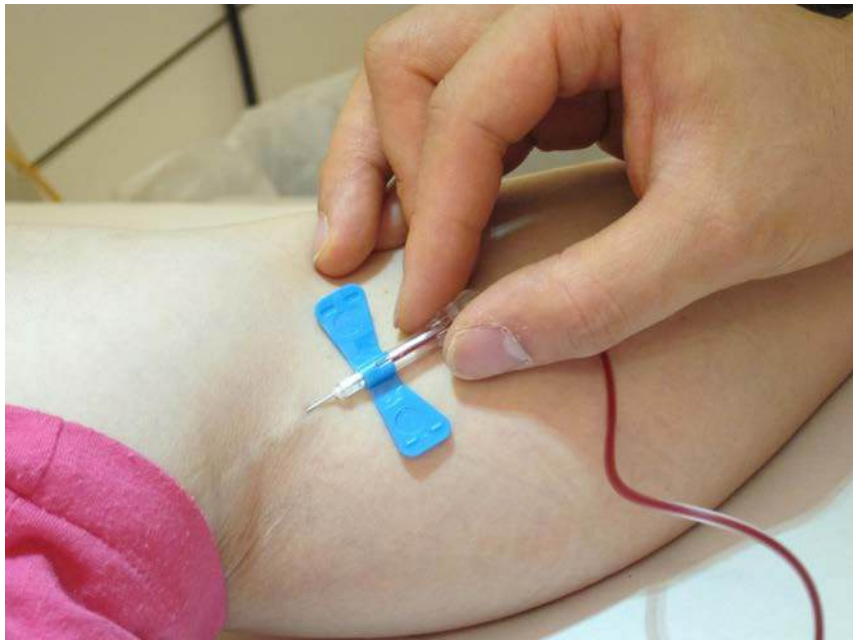
Please show 'Source document 1: Techniques for taking blood from thalidomide-impaired patients' to the person in charge of taking your blood beforehand.

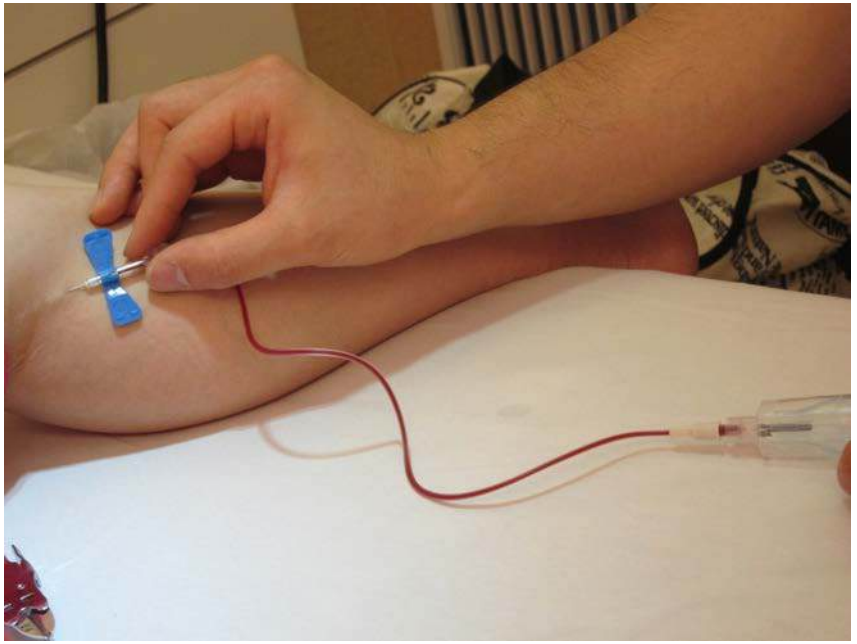
13. Related publications

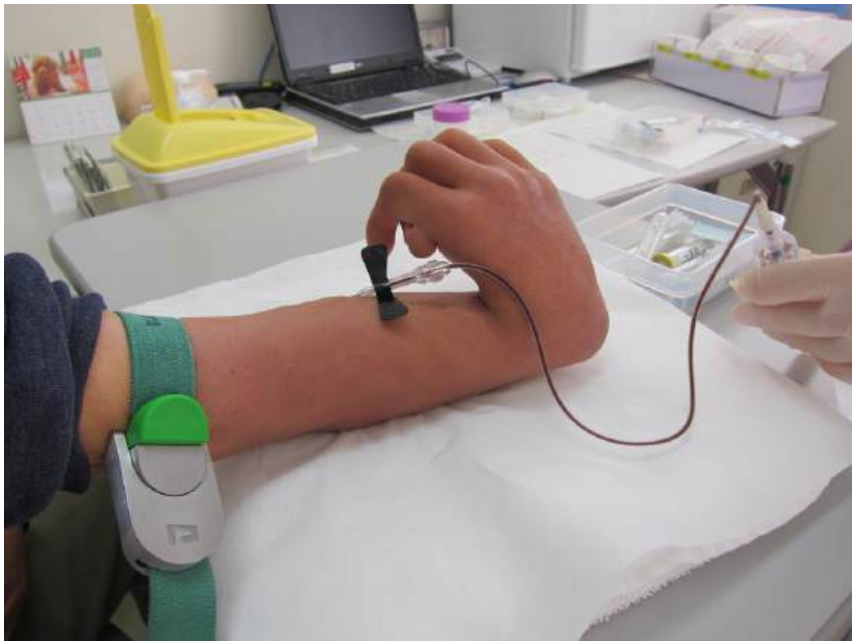
This section is omitted because all related publications are in Japanese.

How to obtain blood samples from thalidomide impaired people

See Q5







*Upper gastrointestinal endoscopy

Throat anesthetic

We will spray something into your mouth.

It has a bitter taste.

Throat anesthetic

Let it sit at the back of your throat for 5 seconds.

Throat anesthetic

Please swallow.

During the test

Please spit out any saliva.

This is to prevent choking.

During the test (breathing)

Breath in through your nose

...and out through your mouth.

Anesthetic

We are going to inject you with a drug.

It will make you feel sleepy.

Relax your neck and shoulders.

The test is finished.

We will return you to your room shortly.

Well done!

You did great.

* Thank you for your cooperation.

When starting the test

We will start the test now

- Thank you for coming today
- My name is …

  and I will be taking care of the tests today
- If you have any questions or concerns,

  please don't hesitate to ask me

Explanation before test

I'll explain the stomach camera test (gastroscopy)

(1) This medicine will get rid of any bubbles in your stomach.
(Please swallow)

(2) Your throat will be numb while the camera is inside you. This means you won't be able to swallow, so please let any saliva dribble out.

(3) We' ll inject some air into your stomach so that we can see it with the wrinkles smoothed out.
Your stomach will feel bloated for about 2 or 3 hours.

(4) You must not eat anything until 1 hour after the test.

Explanation before test

Which nostril is easier to use?

- We'll insert the camera through whichever nostril is larger
- Please press your right nostril and breath in and out through the left nostril
- Now press your left nostril and breath in and out through the right nostril
- Which nostril is more open?

Let's use the

☐ Right ☐ Left

Explanation before test

I'll explain transnasal endoscopy

(1) We'll hold your nostrils open and put some medicine in your nose to prevent it from bleeding.

(Please wait 5 minutes)

(2) We' ll put some anesthetic gel and a tube into your nose to minimize the pain. Please wait 1 minute 30 seconds.

(Swallow any gel that flows down to the back of your throat)

(3) We' ll remove the tube and take you to the gastroscopy room.

(4) The gastroscopy test lasts about 10 to 15 minutes.

Explanation before test

Observations during transnasal endoscopy

We'll look inside your nose, throat and esophagus. Then, we'll move from your stomach to your duodenum, and look as far as where the pancreatic juice and bile are released.

Then we'll return from the duodenum to look closely at the stomach, and we' ll finish by looking again at the esophagus and nasal cavity as we pull the camera out.

Confirmation of identity before test

Now we will do the gastroscopy

- Date of birth:
- Name:

Please check this information.

Is it correct?

☐Yes ☐No

We are going to give you an intramuscular injection

We'll give you an injection to calm down the activity in your stomach

Position during test

Please lie on your left side

(So that you are facing the left)

Precautions during test

Do not swallow your saliva!
You could choke

→ Don't swallow any saliva.
Let it flow out.

How to breathe during the test

…Slowly…

in through the nose,

out through the mouth

During the test

Relax your neck and shoulders

Let yourself be comfortable!

Open your eyes…

Relax your shoulders

Slowly and calmly,

Open your eyes and relax

During the test

Relax even more!

Well done.

You did great!

After the test

That's the end of the test

Explanation of lung function tests

- The first test looks at your lung capacity.
- Hold the mouthpiece between your teeth and breathe through your mouth only.
- When you have the mouthpiece between your teeth, first take a few normal, relaxed breaths.
- When I give the sign, start breathing in and out deeply.

- The next test looks at your lung power when you breathe out.
- First, take a few normal, relaxed breaths, like you did in the first test.
- When I give the sign, breathe in deeply and then breathe out strongly. When you have exhaled all your breath, breathe in deeply one more time.

- The next test looks at how much air is left in your lungs when your are breathing normally.
- Continue breathing in a normal, relaxed way for a little while.
- When I give the sign, breathe in and out deeply.

Breathe in

Breathe out

Explanation of eye tests

(1) Refraction (eye power)

* We will measure your refraction (eye power) with a special instrument.
* You will not feel any pain.
* Please open your eyes wide.

(2) Intraocular pressure (eye hardness)

* We will measure your intraocular pressure (eye hardness) with a special instrument.
* Air will come out of a small hole.
* It will startle you, but it will not hurt.
* Please open your eyes wide.

(3) Visual acuity

* We will measure your visual acuity.
* Please point to the direction of the gap in the circle (C).
* There are four directions – up, down, left, and right.

(4) Photograph of fundus (back of eye)

* We will take a picture of your fundus (back of eye).
* Look at the flashing light inside the instrument.
* Please open your eyes wide.

Gynecological examination

- **This is an examination of your uterus and ovaries.**

(To test for endometriosis, uterine fibroids, cancer, ovarian cysts, etc.)

- **Remove your underwear and sit on the examination table. After sitting, the chair will be raised and you should be lying back with your legs and feet apart. If you relax and breathe through your mouth without tensing your abdomen, the examination will be over quickly and painlessly. Thank you for cooperating.**

- **Cytology: We will examine your cervix to check that there are no polyps or cancerous cells.**

We will insert a metal instrument into your vagina, and gently scrape off and collect mucosal cells using a specially designed instrument.

There will be no pain or bleeding.

Transvaginal ultrasonography: We will insert a narrow probe into your vagina to examine your uterus and ovaries.

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