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?

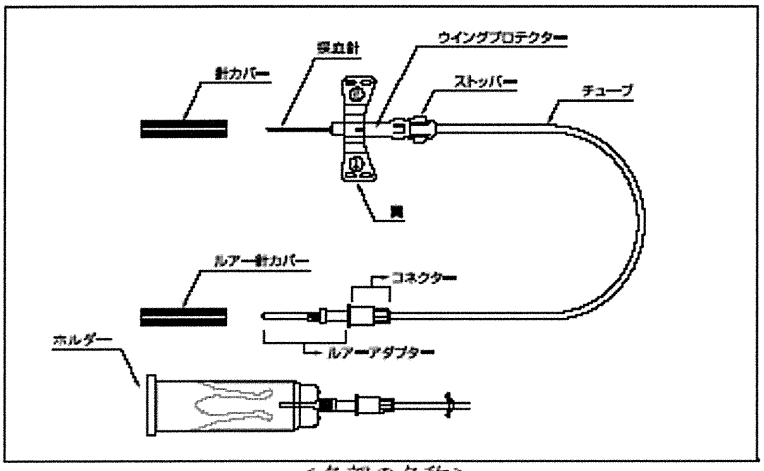
- **See attached source document 1: Techniques for taking blood from thalidomide-impaired patients**

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)

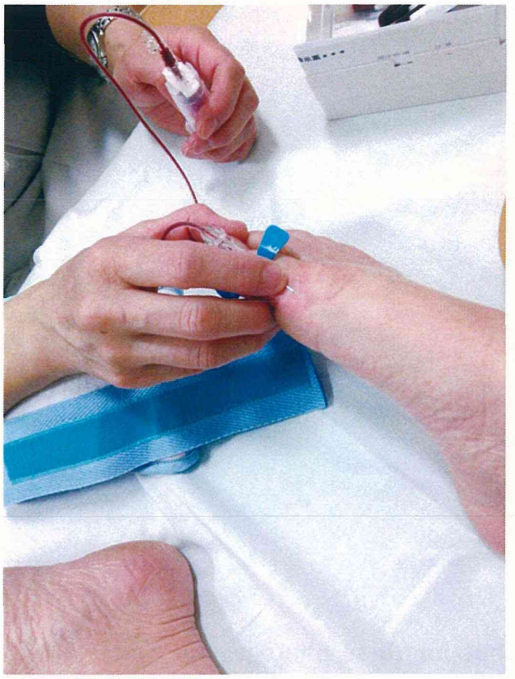


Fig. 2 Blood can only be collected from the medial side of the left knee in this patient (24G)

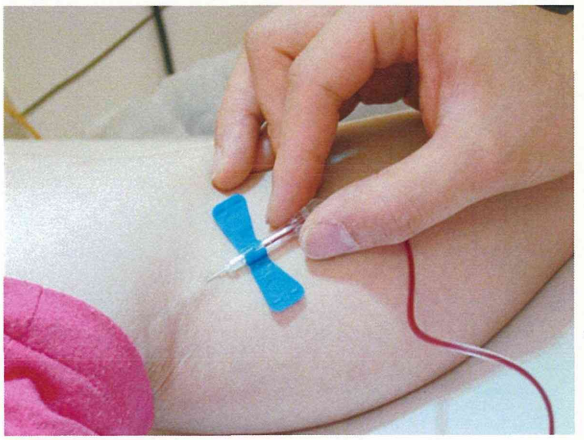
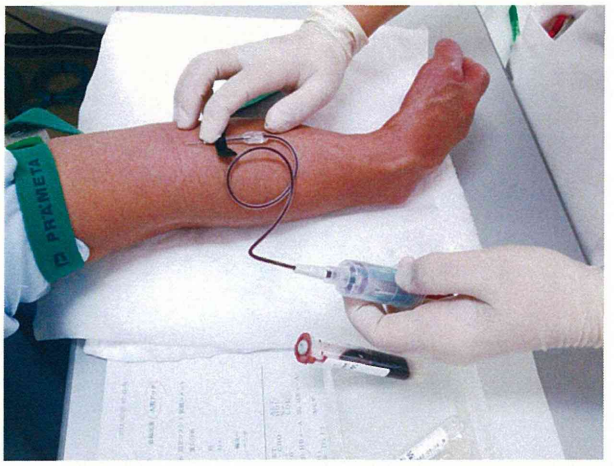


Fig. 3 Blood collection was possible from a cutaneous vein in the right upper limb in this patient (22G)



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.

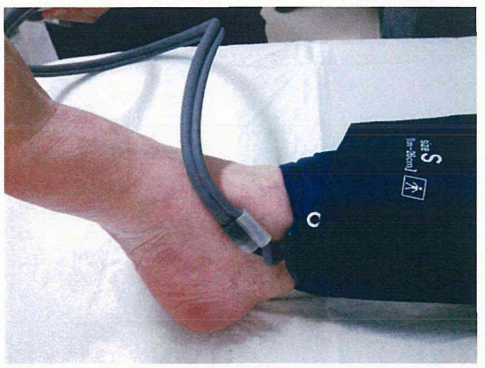


Fig. 1

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

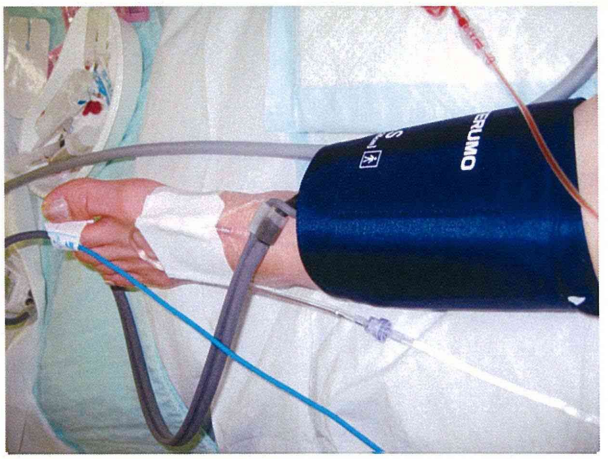
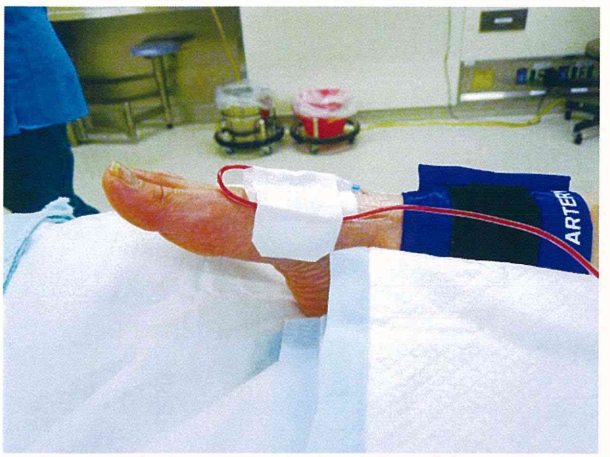


Fig. 4



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.

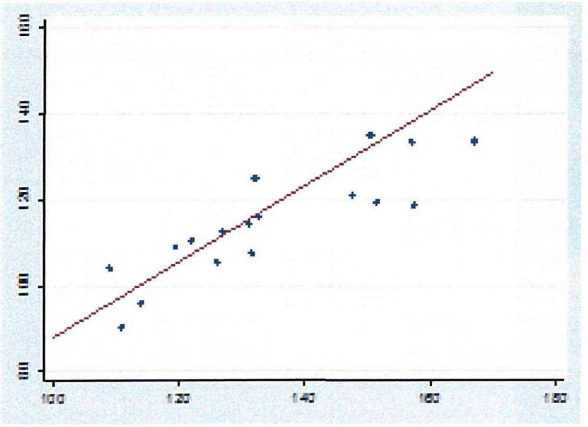


Fig. 5 Relationship between upper and lower limb systolic BP in thalidomide-impaired patients
The horizontal axis shows lower limb BP, the vertical axis shows upper limb BP, and the red line shows the estimation formula 'upper limb systolic BP = 0.88 × lower limb systolic BP'.

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.