

Q&A on Thalidomide-Impaired People

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Ver.5-3

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 $(\text{posterior tibial artery systolic pressure} + 8) \times 0.88$ 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 $SV1 + RV5 \geq 3.5\text{mV}$ or $R5$ or $R6 \geq 2.6\text{mV}$ 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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Table of contents

1. Basic facts about thalidomide impairment

Ryoji Kayamori, Misato Tanaka, Atsuto Yoshizawa

- Q1-1: When did the thalidomide crisis occur and how many Japanese victims are there?
- Q1-2: What types of birth defects did thalidomide cause?
- Q1-3: What are the features of upper limb reduction defects?
- 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?
- Q1-5: What are the important points when examining patients with hearing impairment?
- Q1-6: Are there any impairments other than in the upper limbs and hearing organs?
- Q1-7: Where can I find medical literature on thalidomide?

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

Ryoji Kayamori

- Q2-1 : What are the causes of shoulder pain?
- Q2-2: What causes low back pain?
- Q2-3: What are the main causes of hand numbness?
- Q2-4: What causes persistent pain in the hands?
- Q2-5: What are the causes of hip joint pain?

3. Comments from radiologists

Sayuri Oka, Kazuya Mochiki, Kazuyoshi Yamano,
Toru Sasaki, Tatsuya Wada, Kanehiro Hasuo

- Q3-1: What are the important points when taking X-rays?
- Q3-2: What points should be considered when receiving thalidomide-impaired patients for X-ray?
- Q3-3: What points should be considered when measuring bone mineral density?
- Q3-4: Are there any particular trends in the bone mineral density of thalidomide-impaired patients?

4. Comments from occupational therapists

Takeshi Kobayashi

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

Q&A on Thalidomide-Impaired People

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

Q4-3: I have developed pain in my hip joints and greater difficulty in moving.

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

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.

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.

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

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.

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

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

5. Blood collection

Atsuto Yoshizawa

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

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

6. Measuring and evaluating blood pressure

Yuka Shiga, Yasuhiro Maehara, Atsuto Yoshizawa, Hiroyuki Nagase,

Yutaka Seki, Eriko Kanehisa, Takuro Shinbo

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

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

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?

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

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

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

assistance?

7. Comments from nurses

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

Q7-1: How is blood pressure measured?

Q7-2: How are blood samples obtained?

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

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

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

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

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

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

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

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

8. Upper GI endoscopy (via the mouth)

Toshiyuki Sakurai

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

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

Q8-3: Are sedatives necessary?

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

Q8-5: What sort of endoscope is used?

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

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

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

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

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

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

Q8-12: Are there any other important points?

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

9. Upper GI endoscopy (via the nose)

Takama Maekawa

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

Q&A on Thalidomide-Impaired People

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

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

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

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

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

Q9-7: Are sedatives necessary for transnasal endoscopy?

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

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

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

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

Q9-12: Did any patients have nosebleeds?

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

10. Anesthesia

Yuka Shiga, Yasuhiro Maehara

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

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

Q10-3: Is premedication necessary?

Q10-4: Are any particular preparations necessary?

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

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

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

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

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

11. Psychiatry

Kobun Imai

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

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

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

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

12. Other questions

Atsuto Yoshizawa

- Q12-1: What did you learn from the examination of 76 thalidomide-impaired patients?
- Q12-2: How high does blood pressure measured at the ankle at home have to be for it to be considered hypertension?
- Q12-3: Where is the posterior tibial artery located?
- Q12-4: How do you deal with the fact that blood pressure measured at the lower limbs can differ between left and right?
- Q12-5: Have similar large-scale medical examinations and surveys been done in the United Kingdom and Germany?
- Q12-6: Have other countries also compiled a detailed Q&A on health problems in thalidomide-impaired people?
- 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?
- Q12-8: Do thalidomide-impaired people have any particular health problems to be aware of?
- 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?
- Q12-10: Are people with undeveloped or underdeveloped upper limbs susceptible to gaining weight?
- 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?

13. Source materials

- Source 1: Techniques for withdrawing blood from thalidomide-impaired patients
- 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

1 . Basic facts about thalidomide impairment

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Q1-5: Misato Tanaka

Q1-6: Ryoji Kayamori

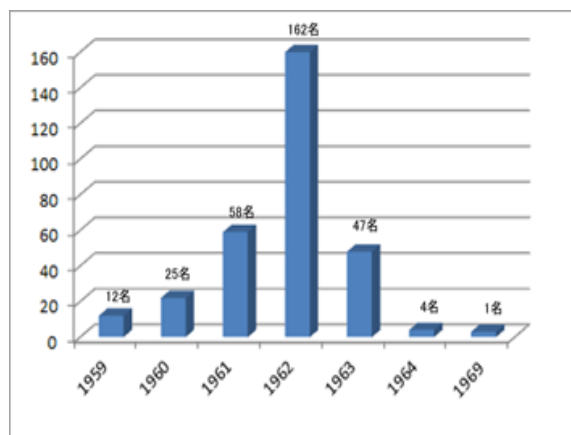
Q1-7: Atsuto Yoshizawa

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?

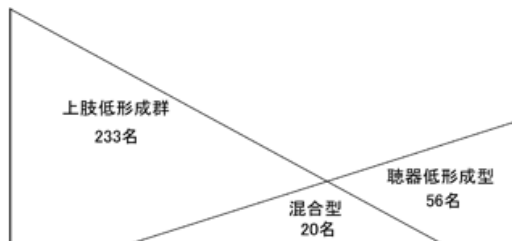
There are three patterns of thalidomide-induced defects: victims either have upper limb reduction defects (also called upper limb phocomelia, upper limb underdevelopment, etc.), hearing organ

1. Basic facts about thalidomide impairment

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?

1. Basic facts about thalidomide 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?

1. Basic facts about thalidomide impairment

- 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)

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|---|----------------|
| • 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 |