The patient survey regarding care of myotonic dystrophy

A. About you (patients)

1. Who is filling out the survey?

(We would like you to answer these survey, but if you cannot respond by yourself, you can ask for help someone to answer.)

- ☐ Patient by yourself
- ☐ Representative
- ☐ Patient with the help of someone

2. How old are you? What is gender?

( )years old

☐Male ☐Female

3. How tall are you? How much do you weight?

Height( )cm、Weight( )kg

4. Where do you live?

( )prefecture

5. Who you live with?

☐Living alone (no housemate)

☐Living with someone: Number of persons excluding you ( )

Number of myotonic dystrophy patients among them ( )

Partner (Spouse) ☐ No ☐ Yes

Children ☐ No ☐ Yes: Number of children ( )

Number of myotonic dystrophy patients among them ( )

Where do you live?

☐Home ☐Institution(NOT hospital) ☐Hospital(Long-term hospitalization)

☐Other( )

Questions for the home care patient. Do you use home service?

☐No(Do not use)

☐Yes : Please, check all home services you use.

☐ visiting doctor ☐ home-visit nursing ☐ Day rehabilitation

☐home-visit rehabilitation ☐home helper ☐guide helper

☐day service ☐home-visit bathing service ☐short stay service

6. Please tell us the allowance or insurance you receive.

Certificate of the Physically Disabled : ☐Grade1 ☐Grade2 ☐over Grade3

Services and Supports for Persons with Disabilities :

☐part1 ☐part2 ☐part3 ☐part4 ☐part5 ☐part6

Long-term care insurance for elderly or specified diseases :

☐Support level1・2 ☐Care Level 1

☐Care Level 2 ☐Care Level 3

☐Care Level 4 ☐Care Level 5

☐Disability pension

☐Special disability allowance

☐Designated intractable disease(Muscular dystrophy)

7. Do you register Remudy (patient registry)?

☐ No ☐ Yes

8. What is your highest educational level? (If you are currently attending a school, please let us know the current situation.)

☐ university • graduate school ☐ junior college ☐ vocational school

high school (☐ general school ☐ for special needs education)

junior high school(☐ general school ☐ school for special needs education)

primary school (☐ general class ☐ special education class ☐ school for special needs education)

☐nursery • kindergarten

9. Please tell about your situation regarding employment.

☐school • under education for employment support

☐community workshop for the disabled

☐employed

working form ;

☐work at home
☐commute to work
commuting method: ☐public transportation、☐own car、
☐transportation service、☐bicycle • on foot
commuting time: about（ ）minutes
☐housewife • help with housework
☐working in the past: retirement age （ ）years old
☐ never worked (excluding people who are in school or under employment support education)

10. Questions for those who answered, “working in the past” or “Never worked” above. What is the reason for retirement or not working?
(Please select all that apply.)
☐mandatory retirement age ☐marriage ☐other personal circumstances exclusive of disease ☐conditions due to DM ☐conditions other than DM
☐others（ ）

B. About your(patients) health

11. How old were you when you first noticed DM-related symptoms?
☐within 4 weeks after birth ☐4 weeks to 1-year-old
☐over 1-year-old: （)years old

12. How old were you when you first visited medical institution for DM or symptoms (complications) related to DM?
（ ）years old ☐unknown

13. What are the symptoms or diseases when you first visit a medical institution?
（ ）

What hospital department did you visit first?
☐neurology • pediatric neurology ☐general internal medicine ☐pediatrics ☐cardiology ☐endocrinology • diabetes ☐gastroenterology ☐respiratory medicine ☐ surgery ☐ ophthalmology ☐ otolaryngology ☐ gynecology

☐dentistry ☐others( )

14. Please tell us about your current walking ability.

☐ambulatory without any support (Not using handrail or stick)

☐able to walk using orthosis (including walker, lower limb orthotic, handrail and stick)

☐abasia: the age when you were unable to walk ( )years old

☐unknown ☐not acquiring a walking ability

15. Do you have the following symptoms? If so, please tell us the imact on daily life and whether receiving any treatment or not.

	No	Yes, but no impact on everyday life	Symptoms are severe and interfere with everyday life / Receiving some kind of treatment
finger impairment (difficult to open a lid etc.)	☐	☐	☐
myotonic phenomenon(difficult to open after gripping hand etc.)	☐	☐	☐
dysphagia (difficult to swallow foods or drinks, foods stick in, etc)	☐	☐	☐
constipation	☐	☐	☐
difficult to see	☐	☐	☐
difficult to hear	☐	☐	☐
fatigue	☐	☐	☐
sleepiness during the day	☐	☐	☐

16. Do you take medicine for myotonia (stiffness)?

☐No

☐Unsure

☐Yes: please list the name of medicine if you know ( )

17. Question for the patient who has dysphagia. Please tell us the age you noticed dysphagia.

( )years old

18. Have you ever been advised to have gastrostomy or tube feeding?

☐ No

☐ Yes : What kind of?

☐gastrostomy ☐tube feeding ☐others( )

Did you have the treatment?

☐Yes : age at which gastrostomy was started ( )years old

age at which tube feeding was started ( )years old

☐No : Why?

☐Did not feel the need. ☐Will have treatment if symptoms become worse. ☐Just want to eat from mouth.

☐Did not want to get invasive treatment.

☐Daily life is restricted. ☐There was no caregiver.

☐Poor physical status. (respiratory function etc)

☐Did not want to receive life extension treatment.

(I think gastrostomy and tube feeding are life-sustaining treatment)

☐others ( )

19. Have you ever had a cataract operation?

☐ No

☐ Yes: age of operation ( )years old

(If you have operations for both eyes, please answer the earlier one.)

20. Do you take medication for fatigue or daytime sleepiness?

☐ No

☐ Yes: please list the name of the medicine if you know ( )

21. Please select all orthoses that you are using.

☐warking stick ☐walker ☐lower limb orthosis

☐manual wheelchairs ☐powered wheelchair ☐electric assisting vehicle

If you are using the following items together with a manual, powered wheelchair, and an electric assisting vehicle, please check.

☐reclining ☐tilt ☐headrest

☐seating systems ☐lift ☐sliding seat

☐self-help devices: ( )

Bathroom equipment: ☐bath chair ☐mat ☐handrail

Restroom equipment: ☐height adjustable toilet seat ☐handrail
☐electric bed ☐special mattress

C. About the hospital where you (patient) get a medical follow-up

22. Do you visit a hospital where the medical staff specializes in neuromuscular disorders? (abbreviated as a specialized hospital below.)

☐Yes. ☐No.

23. Question for those answered "No." in 22.
What is the reason for not visiting a specialized hospital?

☐The specialist/specialized hospital is far from you.
☐I think I do not need a specialist.
☐I did not know that there is a specialist
☐others ( )

24. How long does it take from your house to the specialized hospital you visit? Please select the time to the specialized hospital if you answered specialist doctor/specialized hospital is far from you" in 23.
(Please choose the actual time it requires you to visit the hospital)

☐less than one hour ☐1-3 hours ☐3 hours or more

25. Please select the department that you regularly visit at present. (Including the case of multiple departments in one hospital)

department	At least every half-year	At least once a year	Less than once a year	No visit
neurology pediatric neurology	☐	☐	☐	☐
internal medicine	☐	☐	☐	☐
cardiology	☐	☐	☐	☐
respiratory medicine	☐	☐	☐	☐

Endocrinology / diabetes	☐	☐	☐	☐
gastroenterology	☐	☐	☐	☐
ophthalmology	☐	☐	☐	☐
otolaryngology	☐	☐	☐	☐
dentistry	☐	☐	☐	☐
orthopedics	☐	☐	☐	☐
gynecology	☐	☐	☐	☐
urology	☐	☐	☐	☐

26. Did you have a genetic test for the diagnosis of DM?

☐Yes. When did you receive the test? : ( )years old

☐No.

☐unsure

27. Please select all the tests you received when were diagnosed with DM.

☐electromyogram (Examination performed by inserting a needle in the muscles of limbs)

☐muscle biopsy (Examination to take a part of the muscles of the hands and feet)

☐physical finding (Medical examination such as muscle strength evaluation and myotonic phenomenon)

☐family history (There was DM patient among relatives at the time of consultation)

☐blood test (CK(CPK) was high)

☐others ( )

☐unsure

28. What information and support have you received about your disease so far?

	enough explanation, support	explanation, support insufficient	no explanation and support	unknown
About the clinical course and future	☐	☐	☐	☐

problems				
Information about the disease (such as brochures and websites)	☐	☐	☐	☐
About patient groups	☐	☐	☐	☐
About genetic counseling	☐	☐	☐	☐
About the problem when having a child (infertility, risk of congenital form, etc.)	☐	☐	☐	☐
About psychosocial support (psychological counselor, social worker, introduction of care manager, etc.)	☐	☐	☐	☐
About childcare, education support (cooperation with school)	☐	☐	☐	☐

29. Please check where applicable for the checkups you are receiving and their frequency.

The tests you are receiving	At least every half-year	At least once a year	Less than once a year	Not performed
Muscle strength check	☐	☐	☐	☐
12 lead electrocardiogram	☐	☐	☐	☐
24-hour electrocardiogram (Holter electrocardiogram)	☐	☐	☐	☐
Echocardiography	☐	☐	☐	☐
Respiratory function test (such as vital capacity)	☐	☐	☐	☐
Sleep apnea test	☐	☐	☐	☐

Blood test	☐	☐	☐	☐
Chest radiography (plain x-ray, CT, etc.)	☐	☐	☐	☐
Abdominal echo	☐	☐	☐	☐
Cancer screening	☐	☐	☐	☐
Hearing test	☐	☐	☐	☐

D. About the treatment, you (patients) are receiving

30. Have you ever been advised to use a ventilator etc.?

☐No.

☐Yes. : Do you use ventilator?

☐Yes. Currently using.

☐I had used before but stopped.

☐No. I had never used.

31. Question for the patient who answered "Currently using." in 30.

What kind of ventilation do you use? (Please select all that apply.)

☐non-invasive artificial ventilation during the night or several hours during daytime (assisted ventilation with mask)

☐non-invasive artificial ventilation all-day (assisted ventilation with mask)

☐artificial ventilation by tracheostomy several hours during night or daytime

☐artificial ventilation by tracheostomy all-day

☐mechanical cough assistance (Cough Assist, Pegasus, etc.)

☐manual cough assistance

32. Question for the patient who answered, "I had used before but stopped." in 30.

What was the reason to stop? (Please select all that apply.)

☐wearing a mask was uncomfortable

☐felt uncomfortable for the pressured air

☐felt bloated

☐could not sleep well if I wear a mask

☐difficult to wear and manage at home

☐did not appreciate the effect

☐temporary use in poor physical condition (pneumonia etc.) • stopped with improvement
☐others (　　)

33. Questions for those who answered "No. I had never used." in 30.
What is the reason for not using?
☐Did not feel the need.　☐Daily life would be restricted.
☐There was no caregiver.　☐Too expensive.
☐Did not want to receive life extension therapy.
(I think putting on a respirator is a life-sustaining treatment.)
☐Others　(　　)

34. Have you ever been advised to treat arrhythmia?
☐No.
☐Yes. : What kind of treatment was it?
☐medicine for internal use
☐ablation (cauterize with a catheter)
☐pacemaker implantation
☐implantable defibrillator
☐others　(　　)

35. We ask the patient who answered "Yes." in 34.
Did you receive this treatment?
☐Yes.
☐No.: What is the reason for not receiving the treatment?
☐Did not feel the need.
☐Did not want to receive invasive treatment.
☐Did not want to receive anesthesia.
☐Did not want to receive a catheter treatment.
☐Did not want to use the device.
☐The body condition was bad. (respiratory problem etc.)
☐Others　(　　)

36. Have you ever been advised to treat cardiac dysfunction(heart failure)?
☐No.
☐Yes. : What kind of treatment was it?

☐ myocardial protective agent (ACE inhibitor, angiotensin receptor inhibitor, beta-blocker)

☐diuretics

☐unsure

☐others （ ）

37. Have you ever been advised to treat diabetes/hyperlipidemia (high cholesterol/high triglyceride)?

☐No.

☐Yes. : What kind of treatment was it? (Please select all that apply.)

☐nutrition guidance

☐oral medication

☐Insulin

☐others （ ）

38. Question for those answered "Yes." in 37.

Did you receive the treatment?

☐Yes. : Please select all that apply.

☐nutrition guidance ☐oral medication ☐Insulin

☐No.: What was the reason for not receiving the treatment?

(Please answer also those who have received treatment partially.)

☐Do not feel the need. ☐I want to eat as I like.

☐Bothersome. ☐There is no caregiver. ☐Can not use insulin properly because eyes and hands are impaired.

39. Have you ever had surgery under general anesthesia?

☐No.

☐Yes. How many times? （ ）

Please tell me if you know the details of the surgery. （ ）

40. Question for those who answered "Yes." in 39.

Was the surgery done without any complications?

☐Yes.

☐No.: What kind of complications did you have?

☐delayed arousal from anesthesia ☐extubation difficulty • reintubation

☐rhabdomyolysis • high fever ☐arrhythmia ☐aspiration pneumonia

☐other infections ☐wound healing delay
☐others (　　)

41. Have you ever been treated for infertility?
(You can skip if you do not want to answer)
☐No.
☐Yes. : Did you already have a diagnosis of DM when you received infertility treatment?
☐Yes. ☐No.

42. Do you have an experience of pregnancy, childbirth (please answer only adult female patients)
☐No.
☐Yes. (including miscarriage)

43. Question for those who answered "Yes." in 42.
Where were the pregnancy and childbirth managed?
☐hospital with NICU (neonatal intensive care unit) ☐hospital without NICU
☐clinic ☐midwifery • home

44. Question for those who answered "Yes." in 42.
Were there any complications during pregnancy and perinatal period?
☐No.
☐Yes. : What kind of? (Please select all that apply.)
Pregnancy and perinatal complications
☐amniotic fluid excess ☐asphyxia • respiratory disorder
☐dysphagia ☐hypotonia ☐joint contracture
☐stillbirth and miscarriage ☐unknown
Parturient abnormality
☐cesarean section ☐protracted delivery
☐suction delivery ☐others ☐unknown

45. About rehabilitation
Have you ever been trained on the rehabilitation at home?
☐No. ☐Yes.
Have you been performed rehabilitation by a physical therapist or

occupational therapist?

☐No. ☐Yes. : frequency about ( )a week

Do you receive rehabilitation (such as swallowing, vocalization, higher brain function training) by speech and language therapists?

☐No. ☐Yes. : frequency about ( )a week

46. In the past two years, were there any unexpected hospitalizations by the following reasons?

If there is no hospitalization for the specific reason, please write ″0″.

If there are multiple hospitalizations for the same reason, please indicate the total of all hospitalization days.

in acute respiratory disease (pneumonia, bronchitis, etc) ( )days
heart disease (arrhythmia, heart failure, etc.) ( )days
fractures, injuries, etc. ( )days
gallstones and cholecystitis ( )days
urinary tract infections (such as cystitis) ( )days
diarrhea, enteritis, etc. ( )days
ileus, constipation ( )days
others 〔 〕 ( )days

47. How well do you think your doctor understands your conditions?

☐very understanding
☐quite understanding
☐do not understand much
☐do not understand at all

48. How satisfied are you with your medical treatment?

☐very satisfied
☐quite satisfied
☐not very satisfied
☐not satisfied at all

49. In this survey, is there anything missing regarding the therapy or care you received.
Please feel free to write your opinion or feedback.