European Reference Networks

European Reference Network for rare or low prevalence complex diseases

Network: Neurological Diseases (ERN-RND)

Coordinator: Universitätsklinikum Tübingen — Deutschland

STANDARD OPERATING PROCEDURE (SOP) FOR NERVE CONDUCTION STUDIES IN PATIENTS WITH LEUKODYSTROPHIES AND SUSPECTED PERIPHERAL NERVE INVOLVEMENT

A suggestion for clinical use

to be adapted to local situation

EUROPEAN REFERENCE NETWORKS

FOR RARE, LOW PREVALENCE AND COMPLEX DISEASES

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INTRODUCTION TO THE EUROPEAN REFERENCE NETWORK FOR RARE NEUROLOGICAL DISEASES (ERN-RND)

ERN-RND is a European Reference Network established and approved by the European Union. ERN-RND is a healthcare infrastructure which focuses on rare neurological diseases (RND). The three main pillars of ERN-RND are (i) network of experts and expertise centres, (ii) generation, pooling and dissemination of RND knowledge, and (iii) implementation of e-health to allow the expertise to travel instead of patients and families.

ERN-RND unites 64 of Europe's leading expert centers as well as 4 affiliated partners in 24 member states and includes highly active patient organizations. Centers are located in Austria, Belgium, Bulgaria, Croatia, Cyprus, Czech Republic, Denmark, Estonia, Finland, France, Germany, Greece, Hungary, Ireland, Italy, Latvia, Lithuania, Luxembourg, Malta, Netherlands, Poland, Slovenia, Spain and Sweden.

The following disease groups are covered by ERN-RND:

- Ataxias and Hereditary Spastic Paraplegias
- Atypical Parkinsonism and Genetic Parkinson's Disease
- Dystonia, Paroxysmal Disorder and Neurodegeneration with Brain Iron Accumulation
- Frontotemporal Dementia
- Huntington's Disease and other Chorea
- Leukodystrophies

Specific information about the network, the expert centers and the covered diseases can be found on the network's website ***www.ern-rnd.eu****.*

Recommendation for clinical use:
The European Reference Network for Rare Neurological Diseases strongly recommends the use of the following SOP as best clinical practice for patients with leukodystrophies and suspected peripheral nerve involvement.

DISCLAIMER

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The information (1) should not be considered inclusive of all proper treatments, methods of care, or as a statement of the standard of care; (2) is not continually updated and may not reflect the most recent evidence (new information may emerge between the time information is developed and when it is published or read); (3) addresses only the question(s) specifically identified; (4) does not mandate any particular course of medical care; and (5) is not intended to substitute for the independent professional judgement of the treating provider, as the information does not account for individual variation among patients. In all cases, the selected course of action should be considered by the treating provider in the context of treating the individual patient. Use of the information is voluntary. ERN-RND provided this information on an "as is" basis, and makes no warranty, expressed or implied, regarding the information. ERN-RND specifically disclaims any warranties of merchantability or fitness for a particular use or purpose. ERN-RND assumes no responsibility for any injury or damage to persons or property arising out of or related to any use of this information or for any errors or omissions.

METHODOLOGY

The SOP for NCS was endorsed as recommendation for clinical use by the Disease Group for Leukodystrophies of ERN-RND upon unanimous consent.

Disease group for Leukodystrophies:

Disease group coordinators:

Nicole Wolf[1], Kleopas Kleopa[7], Samuel Gröschel[32]

Disease group members:

Health care professionals:

Marc Engelen[1], Marjo S. van der Knaap[1], Nicola Giannini[2], Leonardo Ulivi[2], Rosa Cortese[3], Odile Boespflug-Tanguy[4], Caroline Sevin[4], Pedro Oliva Nacarino[5], Pierre Kolber[6], Emmanuel Scalais[6], Johanna Uusimaa[9], Hans Hartmann[10], Mar O'Callaghan[11], Beata Błażejewska-Hyżorek[12], Iwona Stępniak[12], Karolina Ziora-Jakutowicz[12], Anna Ardissone[13], Rachele Danti[13], Isabella Moroni[13], Ettore Salsano[13], Duccio Maria Cordelli[14], Veronica Di Pisa[14], Anna Fetta[14], Caterina Garone[14], Luca Solina[14], Susan Byrne[15], Aoife Mahony[15], Pinar Topaloğlu[16], Ángel Martin[17], Thomas Klopstock[18], Ruth Cutajar[19], Sylvia Boesch[20], Lina Schwaighofer[20], Ales Tomek[21], Ieva Glazere[22], Arina Novasa[22], Natalja Predkele[22], Enrico Bertini[23], Giacomo Garone[23], Francesco Nicita[23], Jacopo Sartorelli[23], Gessica Vasco[23], Sabine Grønborg[24], Lena Elisabeth Hjermind[24], Jørgen Erik Nielsen[24], Peter Balicza[25], Laszlo Szpisjak[26], Anneli Kolk[27], Klára Brožová[28], Fran Borovecki[29], Mario Habek[29], Bart Dermaut[30], Dimitri Hemelsoet[30], Tim Rattay[31], Hanna Küpper[32], Lucia Laugwitz[32], Hendrik Rosewich[32], Ludger Schöls[32], Tom de Koning[33], Christa-Caroline Bergner[34], Wolfgang Koehler[34]

Patient representative:

Tomasz Grybek[8], Sara Hunt[8]

Affiliations:

1. *Amsterdam UMC - Amsterdam University Medical Center, Netherlands*.
2. *AOU - University Hospital Pisa, Italy*.
3. *AOU - University Hospital Siena, Italy*.
4. *APHP - Reference Centre for Leukodystrophies, Robert-Debré University Hospital, Paris, France*.
5. *Asturias Central University Hospital, Oviedo, Spain*.
6. *CHL - Luxembourg Hospital Center, Luxembourg*.
7. *Cyprus Institute of Neurology and Genetics, Egkomi, Cyprus*.
8. *ePAG representative*.
9. *Finland Consortium: University Hospitals in Oulu, Tampere and Helsinki, Finland*.
10. *Hannover Medical School, Germany*.
11. *Hospital Clinic Barcelona and Sant Joan de Déu Hospital, Barcelona, Spain*.
12. *Institute of Psychiatry and Neurology, Warsaw, Poland*.
13. *IRCCS - Foundation of the Carlo Besta Neurological Institute, Milan, Italy*.
14. *IRCCS - Institute of Neurological Sciences of Bologna, Italy*.
15. *Irish Consortium: Tallaght University Hospital and Children's Health Ireland*.
16. *Istanbul Faculty of Medicine, Turkey*.
17. *La Paz University Hospital, Madrid, Spain*.
18. *Ludwig Maximilian University Hospital, Munich, Germany*.
19. *Mater Dei Hospital, Msida, Malta*.
20. *Medical University Innsbruck, Austria*.
21. *Motol and Homolka University Hospital, Prague, Czech Republic*.
22. *Pauls Stradins Clinical University Hospital, Riga, Latvia*.
23. *Pediatric Hospital Bambino Gesù, Rome, Italy*.
24. *Rigshospitalet University Hospital Copenhagen, Denmark*.
25. *Semmelweis University, Budapest, Hungary*.
26. *Szent-Györgyi Albert Medical Center, Szeged, Hungary*.
27. *Tartu University Hospital, Estonia*.
28. *Thomayer University Hospital, Prague, Czech Republic*.
29. *University Hospital Center Zagreb, Croatia*.
30. *University Hospital Ghent, Belgium*.
31. *University Hospital Schleswig-Holstein, Kiel, Germany*.
32. *University Hospital Tübingen, Germany*.
33. *University Medical Center Groningen, Netherlands*.
34. *University of Leipzig Medical Center, Germany*.

Endorsement process:

- Proposal to endorse guidelines to Disease Group in terms of annual meeting group discussion – 27 October 2025
- Consent to endorse guidelines by whole Group: 24 June 2026
- Publication of endorsed guidelines: 28 July 2026

LINK TO ORIGINAL PUBLICATION

https://www.mldinitiative.com/wp-content/uploads/2024/08/Nerve-Conduction-Studies-protocol-v1-1.pdf

SOP FOR NCS (AS OF NEXT PAGE)

Source: https://www.mldinitiative.com/
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The MLD initiative

Standard operating procedure

NERVE CONDUCTION STUDIES

(ELECTRONEUROGRAPHY – ENG)

Version	1.0
Status	Definitive version
Date	06/03/2024
Purpose of this document	Describe the standard operating procedure for conducting nerve conduction studies in patients with metachromatic leukodystrophy to enable cross-center comparison and analysis.

Version control

Version number	Revision date	Summary of major changes

A) Baseline parameters

1) Patient information

- Recorded patient information must include: sex, age, body weight, and height

2) Procedural information (please keep constant across measurements)

- Recorded procedural information must include: measurement side, limbs temperature, and exact location of stimulation and recording sites, and use of anesthesia, and type and size of electrode.

3) Source data

- Provide result tables, preferably also curves, graphical views and used normative values

B) Laboratory Procedures

1) Measurement side

- Measurements are always taken on the right side of the body. The left side is optional and is preferably additionally measured in patients with previous investigations on the left side.

2) Type of electrode

- Surface electrodes

3) Parameter Settings according to local lab (please keep constant across measurements)

3.1) Motor Nerve Conduction Studies

Filters:	Low Frequency	according to local lab
	High Frequency	according to local lab
Gain:	0.5 mV – 5 mV/div	

3.2) Sensory Nerve Conduction Studies

Filters:	Low Frequency	according to local lab
	High Frequency	according to local lab
Gain:	1 – 50 μV/div	

Filtersettings according to local lab (add lab)	**Low-freq**	**High-freq**
Motor		
AMC	20 Hz	2 kHz
Tubingen/Milan	20 Hz	5 kHz

Sensory		
AMC	20 Hz	3 kHz
Tubingen/Milano?	20 Hz	2 kHz

3.3) Antidromic

- All sensory studies should be measured in an antidromic direction, except for the medial plantar nerve (only performed in children younger than 12 months of age if sural nerve is not recordable)

4) Temperature Measurements

- The limb temperature will be measured at the beginning and end of each nerve conduction study. The temperature at start and end must be recorded on each nerve tracing.
- For the peroneal, tibial and sural nerve conduction studies, temperature is measured in the lower calf of the side tested. The limbs temperature should be $\geq$ 32°C.

5) Nerve Stimulation

- Surface electrical stimulation will be used. In children under 2 years, we will use a pediatric stimulator with an inter-electrode distance of 15 mm;
- The duration of stimulus current is 0.1 to 0.2 ms, or can be enlarged if necessary (please note)
- Depending on circumstances: often non-recurrent is easier to bare; if recurrent than the stimulation rate is 1/sec.
- The intensity of stimulation should be increased in steps until a supramaximal evoked response is obtained.

6) Averaging

- Sensory responses may be averaged to improve the signal to noise ratio and to facilitate accurate measurement of the onset latency and amplitude.
- The number of averaged responses, up to 10, must be electronically printed out on the tracings

7) Amplitude Measurements

7.1) Motor amplitude

- The CMAP amplitude is measured from the baseline to the negative peak to the nearest 0.1 mV.
- The markers are placed at 500 μV/div; afterwards motor responses may be displayed over 2 divisions for readability.
- Please also provide the area of the negative peak of the CMAP

7.2) Sensory Amplitude

- The SNAP amplitude is measured from the baseline to the negative peak to the nearest 0.1 μV. Please record the measurement used.
- The markers are placed at 10 μV/div.

8) Latency Measurements

- Latencies should be rounded to the nearest 0.1 ms.

8.1) Motor Latency

- The latency is measured at the onset of the negative peak of the CMAP obtained after supramaximal stimulation, or at the onset of the initial positive component if this precedes to negative peak.
- The markers are placed at 500 μV/div.

8.2) Sensory Latency

- The latency is measured at the onset of the negative peak.
- The markers are placed at 10 μV/div.

Optional: Add some figures of electrode placements and marker placements

C) Nerve Conduction Studies in Infants and Children

1) MOTOR STUDY

- The preferred sequence of motor nerve testing is 1) ulnar nerve, 2) peroneal nerve, and 3) tibial nerve (optional).

1.1) ULNAR MOTOR STUDY

Recording site: Abductor digiti minimi (ADM) muscle (medial hypothenar eminence):
G1 placed over the muscle belly
G2 placed over the fifth proximal interphalangeal joint

Stimulation sites: 1. Wrist: medial wrist, adjacent to the flexor carpi ulnaris tendon
2. Elbow: distal to the medial epicondyle

1.2) PERONEAL MOTOR STUDY

Recording sites: Extensor digitorum brevis (EDB) and tibial anterior (TA) muscle:

EDB: G1 placed over the muscle belly
G2 placed distally over the metatarsal-phalangeal joint of the little toe

TA: G1 placed over the muscle belly (1/3 distance fibular head-lateral malleolus
G2 placed 4-6 cm distal at tibia

Stimulation sites:

1. Ankle: anterior ankle, slightly lateral to tibialis anterior tendon
2. Distal to fibular head
3. Proximal to fibular head (optional)

<u>Optional:</u>

1.3) TIBIAL MOTOR STUDY

Recording site: Abductor hallucis brevis (AHB) muscle:
G1 placed over the muscle belly
G2 placed over the metatarsal-phalangeal joint of the great toe

Stimulation sites: 1. Medial ankle: proximal and posterior to the medial malleolus
2. Popliteal fossa: mid-posterior knee over the popliteal pulse

2) SENSORY STUDY

- The preferred sequence of sensory nerve testing is, 1) sural nerve, and 2) median nerve 3) ulnar nerve (optional), 4) medial plantar nerve (only in children younger than 12 months of age if sural nerve is not recordable).

2.1) ULNAR SENSORY STUDY (ANTIDROMIC)

Recording site: V^{th} finger (digit 2):
Ring electrodes with G1 placed over the metacarpal-phalangeal joint
G2 placed 2-3 cm distally over the distal interphalangeal joint

Stimulation sites: 1. Wrist: medial wrist, adjacent to the flexor carpi ulnaris tendon
2. Elbow: at the medial epicondyle

2.2) SURAL SENSORY STUDY (ANTIDROMIC)

Recording site: posterior to the lateral malleolus
G1 placed posterior to the lateral malleolus
G2 placed 2 cm distally
Average up to 10 responses

Stimulation site: Over the sural nerve (4-10 cm proximal to the ankle)

Optional:

2.3) MEDIAN SENSORY STUDY (ANTIDROMIC)

Recording site: Index finger (digit 2):
Ring electrodes with G1 placed over the metacarpal-phalangeal joint
G2 placed 2-3 cm distally over the distal interphalangeal joint

Stimulation sites: Wrist: middle of the wrist between the tendons to the flexor carpi radialis and palmaris longus
Antecubital fossa: medial to the brachial artery pulse

2.4) MEDIAL PLANTAR (ORTHODROMIC) only in children younger than 12 months of age

Recording Site: Medial ankle:
G1 placed slightly proximal and posterior to the medial malleolus
G2 placed 3–4 cm proximally

Stimulation Sites: Great toe (medial plantar sensory):
Ring electrodes, with cathode placed proximally near the metatarsal-phalangeal joint of the great toe; anode placed 1-1.5 cm distally

Optional: Add some figures of electrode placements and marker placements

European Reference Networks

https://ec.europa.eu/health/ern_en

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Network
Neurological Diseases
(ERN-RND)

Coordinator
Universitätsklinikum
Tübingen — Deutschland

www.ern-rnd.eu

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