allowing physical examination after adequate triaging in selected patients. Although, rural India is poor in individual digital literacy, there is a wide network of e-mitra kiosks, ASHA workers and teachers who have come forward to help navigate the system and move through the process. The benefit arising out of limited physical visits to the hospital for patient are already described but restricting the exposure of doctors and patients to someone who is potentially infected is of vital importance.

For a major impact to be seen, an operational telehealth network is required, and infrastructure needs to be scaled up. It also requires a behavior change of not just an individual or an institute but an entire health system as well as patients. We have tried to curtail these limitations and made a beginning while making use of the COVID-19 crises as an opportunity to introduce the system that will stay for future.

Acknowledgements: Prof Kuldeep Singh, Dean Academics and Prof Sanjeev Misra, Director, AIIMS, Jodhpur for conceptualizing the idea and facilitating the development of the software.
Published online: August 11, 2020; *PII*: S097475591600229

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REFERENCES

1. Mahajan V, Singh T, Azad C. Using telemedicine during the COVID-19 pandemic. Indian Pediatr. 2020;57:652-7.
2. Ministry of Health and Family welfare, Government of India. Telemedicine Practice Guidelines Enabling Registered Medical Practitioners to Provide Healthcare Using Tele-medicine. New Delhi. Available at *https://www.mohfw.gov. in/pdf/Telemedicine.pdf.* Accessed on April 03, 2020
3. Smith AC, Thomas E, Snoswell CL, *et al*. Telehealth for global emergencies: Implications for coronavirus disease 2019 (COVID-19) [published online ahead of print, 2020 Mar 20]. J Telemed Telecare. 2020;1357633X20916567.
4. Tolone S, Gambardella C, Brusciano L, G del Genio, Lucido FS, Docimo L. Telephonic triage before surgical ward admission and telemedicine during COVID-19 outbreak in Italy. Effective and easy procedures to reduce in-hospital positivity. Int J Surg. 2020; 78:123-25
5. Hollander JE, Carr BG. Virtually perfect? Telemedicine

Proximal Limb Girdle Weakness, Joint Hyperlaxity, and preserved Deep Tendon Reflexes: A Distinctive Phenotype

A 9-year-old girl presented with mild motor delay and progressive proximal limb-girdle weakness. Socio-cognitive milestones were normally attained. Examination revealed normal head size and intellectual functioning, proximal limb girdle weakness, mildly prominent calves, and preserved deep tendon jerks (including both ankles). She had hyperlaxity of finger joints and both elbow joints (Beighton score 4/9). She also had polyminimyoclonus. Creatine kinase levels were elevated (790 IU/L) while electrocardiogram revealed tremor (***Fig.* 1**). Nerve conduction studies revealed motor axonal loss with sensory sparing while electromyography (EMG) was suggestive of abnormal spontaneous activity (fibrillations and fasciculations) signifying active denervation. She was not cooperative for voluntary EMG assessment. Multiplex ligation-dependent probe amplification (MLPA) revealed homozygous deletion of exon 7 and 8 of *SMN1* gene confirming the diagnosis of spinal muscular atrophy type 3 (SMA type 3).

Important differential diagnosis for progressive limb girdle weakness presenting in late childhood (with onset beyond infancy) include muscular dystrophies (especially Duchenne muscular dystrophy and limb girdle muscular dystrophies) and SMA type 3. It may be difficult to differentiate these conditions based on deep tendon jerks and creatine kinase levels because these are often misleading. Deep tendon reflexes may be preserved in SMA type 3 [1]. Joint hypermobility and hyperlaxity, although an overlooked feature of SMA, if present favors a diagnosis of SMA over muscular dystrophy [2,3]. The caveats include early-onset muscle disorders such as congenital muscular dystrophies and congenital myopathies [2]. In SMA,

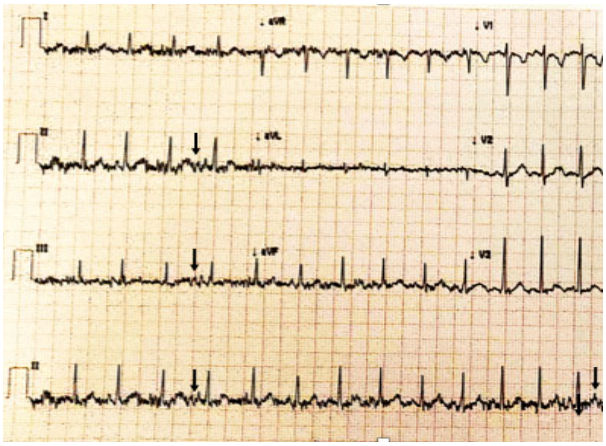

Fig. 1 *Electrocardiogram of the index patient showing the high frequency (30-40 Hz) tremor (arrows) due to muscle fasciculations (seen predominantly in limb leads).*

anterior horn cell loss begins in early infancy and may possibly account for distal hypotonia and hyperlaxity. Hyperlaxity, especially of upper limb joints may persist till adulthood in more than half of patients [4]. It is perplexing to see that this finding was not captured in major prospective cohorts of SMA type 2 and 3, which predominantly addressed the weakness and ambulation. This finding needs to be further confirmed in large cohorts not only because of diagnostic significance but also for rehabilitation point of view, considering the improved outcomes with newer therapies in SMA.

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REFERENCES

1. Lannaccone ST, Browne RH, Samaha FJ, Buncher CR. Prospective study of spinal muscular atrophy before age 6 years. Pediatr Neurol. 1993;9*:* 187-93.
2. Donkervoort S, Bonnemann CG, Loeys B, Jungbluth H, Voermans NC. The neuromuscular differential diagnosis of joint hypermobility. Am J Med Genet C Semin Med Genet. 2015;169C:23-42.
3. Haaker G, Fujak A. Proximal spinal muscular atrophy: Current orthopedic perspective. Appl Clin Genet. 2013;6:113-20.
4. Tofts LJ, Elliott EJ, Munns C, Pacey V, Sillence DO. The differential diagnosis of children with joint hypermobility: A review of the literature. Pediatr Rheumatol Online J. 2009;7:1.

NeoBox - A Multipurpose Aerosol Box for Neonatal Care DuringCOVID-19 Pandemic

Safety of the newborn and the protection of healthcare workers (HCWs) from aerosol exposure are extremely important during the current severe acute respiratory illness coronavirus 2 (SARS-CoV-2) pandemic. Use of personal protective equipment (PPE) has been shown to be associated with a reduced risk of infection [1]. As per WHO guidelines, it is mandatory to use personal protective equipment (PPE) while performing aerosol-generating procedures like suction, intubation, chest compression *etc*. [2,3]**.** However, despite the use of PPE, there remains a possibility that aerosols can contaminate nearby surfaces [4]**.** An aerosol box acts as a physical barrier against the aerosol spread [4,5]. PPE with a barrier enclosure like an aerosol box can be an effective measure to minimize aerosol spread and exposure during this pandemic [2,4].

Recent literature reports that when an aerosol box was used for airway management, the inner surface of the box and the laryngoscopist's gloves and gowned forearms were observed to be contaminated [4], but no macroscopic contamination outside the box was observed [4]. Unlike adult resuscitation, the focus of newborn resuscitation is effective ventilation of baby's lungs which includes aerosol-generating procedures like suction, PPV, using continuous positive airway pressure, intubation, chest compression, etc. [6,4]. To see the feasibility of using the standard aerosol box as a barrier enclosure while performing aerosol-generating procedures on neonates, a novel simulation session integrating a newborn delivery of a suspect or confirmed COVID-19 mother with a subsequent need for neonatal resuscitation was developed. The 15-minute simulation was run with two resident doctors, an embedded simulation nurse, and a low fidelity manikin in the delivery room setting. The 'newborn' was a low fidelity simulator (Laerdal Medical). During simulation sessions, accessing the neonate and performing resuscitation steps in the squared aerosol box was observed to be impossible. After completion of each session, the learners were debriefed using the PEARLS Healthcare Debriefing Tool with plus/delta and advocacy enquiry format by a trained simulation leader [8,9]**.** Difficulties were encountered at all steps of resuscitation like - attaching pulse oximeter, performing positive pressure ventilation, intubation, chest compression and umbilical catheterization, etc. These difficulties were addressed and the need for a modified aerosol box for neonates was informed to the biomedical department of our institute. The box underwent multiple modifications based on the feedbacks received. The final design specifications were given (***Web Table* I**) and the NeoBox was developed (***Fig.*1**).

The NeoBox is made up of a transparent polycarbonate (3 mm thick). The material required was procured and necessary fabrications were done by the local acrylic / polycarbonate sheet fabricator. The average time required to make one NeoBox was approximately 4 hours. The cost was Rs 6500. An alcohol based disinfectant (Ethanol 70%) with a contact time of minimum 1 minute is used to clean the NeoBox [10].

The NeoBox was primarily designed as a physical barrier to prevent aerosol exposure and spread while performing aerosol-generating procedures during resuscitation in delivery room. While running simulation sessions, its wider application for neonatal care like transporting a suspected or confirmed COVID-19 neonate from one place to another (intra hospital transport) and caring for them in the neonatal intensive care unit (NICU) while performing aerosol-generating procedures was recognized. Use of NeoBox in addition to PPE helped boosting HCWs confidence for managing suspected or confirmed COVID-19 neonates. We found that the NeoBox would require training before use in the treatment of patients. Wearing PPE is