Titinopathy – a rare cause of multiorgan failure

337 DEPARTMENT OF PEDIATRICS, CLINICAL HOSPITAL CENTRE ZAGREB

Ivana Čičak*, ¹Milivoj Novak, ¹Miran Cvitković, ¹Slobodan Galić, ¹Toni Matić, ¹Filip Rubić, ^{1,4}Ivan Malčić, ¹Dalibor Šarić, ¹Dorotea Bartoniček, ^{1,4}Daniel Dilber, ²Dražen Belina, ¹Vesna Benjak, ¹Andrea Dasović-Buljević, ^{1,4}Ruža Grizelj, ^{1,4}Boris Filipović-Grčić, ¹Dorotea Ninković, ^{1,4}Jurica Vuković, ¹Lana Omerza, ¹Mirna Aničić, ^{3,4}Anko Antabak, ^{1,4}Mario Čuk. ¹Department of Pediatrics; ²Cardiac Surgery, Clinical Hospital Centre Zagreb; ³Pediatric Surgery, Clinical Hospital Centre Zagreb; ⁴School of Medicine, University of Zagreb

10.1136/archdischild-2021-europaediatrics.337

We report the case of a full term male child with multiple organ dysfunctions caused by mutations of the TTN gene. Our patient had cardiac abnormalities verified by fetal echocardiography at 26 weeks of gestational age, when fetal hydrops due to cardiac decompensation was shown.

Due to specific appearance seen at birth – generalized oedema, light facial and sexual dimorphism with hypogenitalism and chryptorchidism, axial hypotonia in addition to the proven congenital heart defects, a genetic disorder or syndrome with muscle involvement was suspected. Multiple comorbidities were present since birth, mainly cardiac and neurologic, some of which have been described in patients with core myopathies (atrial and ventricular septal defects, supraventricular tachycardia, hypotonia, developmental delay, myopathy). Due to the complexity of the heart defect at the age of 2 months a partial pulmonary trunk banding and ligation of the ductus Botalli (PDA) was initially carried out, and at 9 months a complete correction of heart defect was performed. Our patient also had failure to thrive as well as recurrent vomiting, which was later on associated with extraluminal compression of the proximal duodenum, intestinal malrotation and malfixation. Although a surgical approach somewhat increased tolerance of peroral food intake, chronic diarrhoea and failure to thrive persisted.

In addition, our patient had several metabolic and endocrine disorders including hypocalcemia, hypomagnesemia, hyponatremia, repeatedly elevated levels of PTH, hypothyroidism etc. Our patient had recurrent, mainly respiratory infections with multiresistant microorganisms, and required intubation and long-lasting mechanical ventilation. The above mentioned multiple disorders of various organ systems, with complications caused by multiple sepses, ultimately resulted in multiorgan failure, and despite all the applied treatment at the ICU, resulted in a lethal outcome. Corresponding findings described in other patients with core myopathies included neonatal hypotonia, poor suckling, severe motor skill delay, complex heart defects and cardiomyopathy.

338 LYELL'S SYNDROME – POTENTIALLY LIFE-THREATENING MUCOCUTANEOUS DISEASE

Marko Bašković*, Zoran Barčot, Dora Škrliak Šoša, Davor Ježek, Božidar Župančić. *Children's Hospital Zagreb*

10.1136/archdischild-2021-europaediatrics.338

A five year-old girl came to the tourist ambulance because of the dysuria, sore throat and tingling of external genitalia. A

local physician prescribed amoxicillin/clavulanic acid suspension. She took her first dose in the afternoon. During the night, parents noticed the rash and redness of the entire body. During the night, another dose of antibiotic was given, after which the rash and redness continued to spread rapidly.

Immediately methylprednisolone and chlorpyramine are administered intramuscularly. Upon arrival to the hospital, the girl was a normal state of consciousness with dyspnea, breathing frequency 25/min, SpO₂ 97%, dehydrated, febrile 38.1 °C, tachycardic (135/min) and normotensive (RR 103/66 mmHg). On the skin of the face and on the larger surface of the body (TBSA = 80%) were visible bullae and vesicles which ruptured and it came to skin peeling. Due to the progression of respiratory insufficiency, tracheotomy was performed. Also due to the increase in inflammatory parameters for the first ten days, ceftriaxone and amikacin were ordered, with all supportive therapy. On the second day of admission, cyclosporine is ordered. All the swabs were negative. Lyell's syndrome, or toxic epidermal necrolysis, is a rare, potentially life-threatening mucocutaneous disease, usually provoked by the administration of a drug and characterized by acute necrosis of the epidermis. The drugs most frequently incriminated are nonsteroidal antiinflammatory drugs, chemotherapeutics, antibiotics, and anticonvulsants. Although the cases where amoxicillin/clavulanic acid suspension caused this condition were described, in our case, amoxicillin/clavulanic acid suspension was probably not the cause, given that the girl had been treated twice in the past with this drug without side effects.

339 DEVELOPING A NOVEL MULTI-DISCIPLINARY IN-SITU SIMULATION EDUCATION PROGRAMME FOR PAEDIATRIC EMERGENCY TRAINING FOR NCHDS, AT UNIVERSITY HOSPITAL GALWAY, IRELAND

Aoife Flynn*, Marcus Jee, Jamie Davis, Daniel Khamoudes, Ethel Ryan, Elaine Reade. *Paediatric Department, University Hospital Galway*

10.1136/archdischild-2021-europaediatrics.339

Introduction In the Emergency Department at University Hospital Galway, a tertiary teaching hospital in Ireland, we observed that the Paediatric team predominantly managed medical emergencies while our Emergency Medicine colleagues managed only surgical/trauma cases. Therefore, we developed a novel interdisciplinary education programme that utilises in-situ simulation to enhance training in management of Paediatric emergencies by NCHDs.

Aims In response to preparing NCHDs for a variety of Paediatric emergency scenarios, our programme aimed to share insights through practice and enhance the simulated education experience; by standardising the approach using a pre-brief/debrief format, and improving interdisciplinary knowledge, communication and engagement, through preparation, role allocation and pre-briefing for participants.

Methods We conducted a series of interdisciplinary, in-situ simulations which involved a multidisciplinary response to a medical/surgical case. The scenarios were written by Specialist Registrars from both teams, with a standardised format/template. We implemented a weekly rota whereby the Paediatric team ran the surgical/trauma based cases and the ED team ran the medical based emergencies, with facilitation by Specialist Registrars, supervision by both ED and Paediatric Consultants.

Participants, who were pre-selected, were privy to scenarios/resources/guidelines beforehand.

Considering restrictions, due to COVID19 outbreak, we implemented video sessions to reduce the number of observers present while extending education to the full complement of both faculties.

Results Based on pre/post-simulation questionnaires, over 70% of participants reported the simulations improved their skillset and they felt more confident in participating in future; 100% now felt better equipped to manage Paediatric Emergencies. Engagement in the video streaming was most encouraging, in an era of social distancing and virtual based education.

Conclusion In conclusion, implementing a novel approach to an innovative and multidisciplinary education programme, through the modality of in-situ simulation, can enhance the learning experience and broaden the knowledge base, of NCHDs. We hope that our experience may be used as foresight by others in developing their own multidisciplinary in-situ programme.

340

ETHICAL CONTENT OF GUIDELINES FOR END-OF-LIFE DECISION-MAKING IN PEDIATRIC AND NEONATAL INTENSIVE CARE UNITS: A SYSTEMATIC REVIEW

Sunčana Janković*, Dina Vrkić, Marko Čurković, Bojana Nevajdić, Milivoj Novak, Štefan Grosek, Chris Gastmans, Bert Gordijn, Branka Polić, Ana Borošević. *University hospital center Zagreb, Croatia*

10.1136/archdischild-2021-europaediatrics.340

The aim of this study was to search for all available reviews, guidelines and analyses and make a literature overview and comparison between countries as a corner stone for a further research with a goal of making our national pediatric and neonatal end-of-life guidelines.

We conducted a literature search in February 2018 and in September 2019 in bibliographic databases and grey literature sources for the time period from 1990 to 2019. Search strategies in were conducted using MeSH terms and keywords related to 'pediatric', 'neonatal', 'guidelines', 'end of life', 'palliative care' and 'intensive care unit' terms. Only documents satisfying all of the inclusion and exclusion criteria were included in the review. This resulted in 12 eligible documents.

Ten papers talk about neonatal and four about pediatric issues. Throughout all analyzed papers all of the ethical principles and dilemmas have been mentioned. Best interest model and judicial clarification when parental and physician views collide; doctrines of double effect and omission for withholding and withdrawing of treatment; active and passive euthanasia; quality of life from patient's/parent's and physician's point of view; autonomy and autonomy by proxy; veracity as an informed consent due to shared decision making process; professional duty; beneficence; compassion and nonmaleficence for alleviate suffering; transparency as concealing medical records or documenting detailed plans of care; equality, fairness, social justice and proportionality through treatment justification and allocation of resources for expensive process of treatment or collective society ethics to prevent creation of handicaps; efficacy; utility.

By many means pediatric population is very specific as well as relationship that pediatricians build with children as patients and their parents. Death after forgoing life-sustaining-

treatment in intensive care unit occurs through a procedure conforming to national ethical guidelines which in turn seem appropriate for newborn infants and children. Subjectivity regarding indirect euthanasia seems unavoidable. Systematic teaching of ethics to all intensive care staff and continued review processes of end-of-life situations are necessary to preserve the best interests of the critically ill children and relieve their families. The degree of involvement of the parents in the decision-making process varies according to cultural factors and to the structure and functioning of the medical team in charge.

*This article is part of the research 'Values and Decisions at the End of Life', funded by Croatian Science Foundation.

341

CLINICAL CONTENT OF GUIDELINES FOR END-OF-LIFE DECISION-MAKING IN PEDIATRIC AND NEONATAL INTENSIVE CARE UNITS: A SYSTEMATIC REVIEW

Sunčana Janković*, Dina Vrkić, Marko Čurković, Antonija Janković, Milivoj Novak, Štefan Grosek, Chris Gastmans, Bert Gordijn, Branka Polić, Ana Borošević. *University hospital center Zagreb, Croatia*

10.1136/archdischild-2021-europaediatrics.341

The aim of this study was to search for all available reviews, guidelines and analyses and make a literature overview and comparison between countries as a corner stone for a further research with a goal of making our national pediatric and neonatal end-of-life guidelines.

We conducted a literature search in February 2018 and in September 2019 in bibliographic databases and grey literature sources for the time period from 1990 to 2019. Search strategies in were conducted using MeSH terms and keywords related to 'pediatric', 'neonatal', 'guidelines', 'end of life', 'palliative care' and 'intensive care unit' terms. Only documents satisfying all of the inclusion and exclusion criteria were included in the review. This resulted in 11 eligible documents.

Most of the articles, eight of them are focused on neonatal issues and only three mention specificity of pediatric problems. Among all articles five are general guidelines with examples of diagnosis, five have week by week algorithm and classification of neonatal interventions and three have obstetrical algorithms of prenatal interventions, one article talks about perinatal prognostic factors, four about starting provisional intensive care and reassessment when in doubtful cases, six have a list of clinical procedures in end-of-life decisions, four articles point out a prevalence of neurological impairment after survival as a relevant clinical factor in future end-of-life decisions, four tackle with question of active euthanasia, two recommend palliative care after forgoing life-sustaining treatment with examples of diagnosis, three distinguish different modes of forgoing life-sustaining treatment and two mention organ donation after death.

Clinical guidelines on EOLD are intended as a general framework to help all decision-making parties, but because of uniqueness of every physician, parent, child, diagnosis and underlying background there are still grey zones that tackle with ethical issues and individual decisions outside the guidelines are always possible.

*This article is part of the research 'Values and Decisions at the End of Life', funded by Croatian Science Foundation.