

OrphanAnesthesia Project: New emergency card to augment recommendations for anaesthesiological management in the presences of rare diseases

C. Gaik¹ · T. Münster²

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Summary

For a number of years, the internet platform “OrphanAnesthesia” has provided recommendations for the anaesthesiological management of patients suffering from rare diseases. Going forward, an “emergency card” aimed at providing a systematic overview of important information to support anaesthesiologists providing urgent care for such patients will form a part of all new recommendations, whilst also being added to existing recommendations down the line. The emergency card was developed following the principles of the internationally familiar ABCDE approach to prioritising care for critically ill or injured patients. The modified approach intentionally maintains the prioritisation from “A” to “E” in relation to anaesthesiological workflows, making relevant information on urgent care in the presence of a rare disease readily available, thereby increasing patient safety.

The project „OrphanAnesthesia“

The internet platform “OrphanAnesthesia” publishes information about anaesthesia in cases of rare diseases. The project was launched in 2011 with the aim of increasing patient safety by issuing action recommendations for the narcosis of patients suffering from a rare disease. OrphanAnesthesia was initiated by the Science Working Group of the German Society of Anaesthesiology and Intensive Care Medicine (Deutsche Gesellschaft

für Anästhesiologie und Intensivmedizin, e. V. DGAI) in cooperation with Orphanet. International anaesthetists and disease experts have been creating and regularly revising new recommendations for action in order to increase the safety of anaesthesiological management in cases involving rare diseases [2,3].

The planning, performance and special situations likely to occur during the anaesthesia of patients inflicted with rare diseases and syndromes are explained in a structured manner and substantiated with current medically relevant data and case reports.

The platform combines the expertise gained from a multitude of scientific publications and the experiences of certified experts. “OrphanAnesthesia” then makes the anaesthesia-related information accessible to physicians, patients and self-help organisations free of charge, worldwide and in various languages [3].

A patient card similar to the anaesthesia pass

All recommendations are subject to an international peer review process in which an anaesthetist as well as an additional disease expert take part [3]. In addition, “OrphanAnesthesia” provides patients with a card which is similar to the anaesthesia pass, stating the patient’s rare disease and referring to the “OrphanAnesthesia” platform (Fig. 1) [4].

- 1 Klinik für Anästhesie und Intensivtherapie, Universitätsklinikum Marburg (Direktor: Prof. Dr. M. Wulf)
- 2 Klinik für Anästhesie und operative Intensivmedizin, Krankenhaus Barmherzige Brüder Regensburg (Chefarzt: Prof. Dr. T. Münster)

Competing interests

The authors declare no competing interests.

Keywords

Anaesthetics and Rare Diseases
– Rare Disease – Emergency Information in Anaesthesia
– Orphan Disease – Orphan Anaesthesia – Emergency Card – Syndromic Patients

Figure 1



The name of the respective rare disease shall be entered on the emergency card. The card can be downloaded and printed out at www.orphananesthesia.eu. In the best possible case, it should be submitted to the anaesthetist prior to anaesthesia. Ideally, patients should carry the emergency card with them at all times in order to be prepared for emergencies. By accessing the OrphanAnesthesia platform, the treatment team will then be able to obtain more information about the anaesthesiological requirements of the patients concerned [4].

Anaesthetists are able to read important information either at the premedication outpatient department or perioperatively. The patient pass and/or patient card are to make a contribution towards increased patient safety of this special clientele and will be able to function as an interface between those affected and the anaesthesia team.

Not much time, only few data available – a common problem in an emergency situation

As a rule, in most cases there is only little corroborated available knowledge pertaining to a rare disease, and even less on potential special features to be observed when anaesthesia is performed [3]. For this reason, many recommendations rely on case reports, as applicable guidelines or algorithms do not exist, due to the sometimes very low numbers of cases of the disease. Proposals for a structured approach for anaesthesiological preparations in case of rare diseases are already available [1, 5]. In the event of unexpected complications and in life-threatening emergency situations (in-hospital and emergency medical service), however, there will often be no time for reading these publications or for a detailed study of potentially applicable recommendations

In addition, when treating patients with rare diseases, particularly anaesthetists at smaller hospitals often do not have the opportunity to consult colleagues

representing other specialties (e.g. neurology or paediatrics) for advice. If the rare disease itself results in an indication for surgery or intervention, in the best case the surgeon will be able to provide valuable information prior to surgery. It is particularly the combination of “little time” and “scarce data” that might correlate inversely with the potential hazard of patients suffering from a rare disease while receiving anaesthesiological care.

In order to give the attending anaesthetists in charge of treatment a systematic overview including the most important emergency situations even in time-pressing situations, an “emergency card” shall now become a part of the existing and future recommendations for action. In the future, if a patient is known to have a rare disease and a corresponding recommendation for action is available at “OrphanAnesthesia”, the emergency information referring to anaesthesiological key topics such as airway, blood circulation and/or haemodynamics, transfusion management and can be immediately viewed. The emergency card will soon contain all new and updated recommendations for action and recommendations already published will be added to the respective emergency cards successively in the course of time.

For anaesthesia and sedation in cases of rare diseases, Butler et al. developed a checklist containing the items “Difficult Airway”, “Respiratory Pathologies”, “Gastral Reflux”, “Cardiovascular Disorders”, “Neuromuscular Problems”, “Function-

nal Disorders of Liver and Kidneys” [1]. The (potentially) difficult airway, pulmonary/respiratory pathologies and a gastroesophageal problem will be represented by the letter “A” and/or the category of “Airway/Anaesthetic Technique”. Due to their relevance in an anaesthesiological setting, “Blood circulation and Haemodynamics” constitute a category of their own. The same applies to the items “Coagulation/Blood Products”. In analogy to Butler et al., the OrphanAnesthesia emergency card registers kidney and liver function via the columns “Drugs” (e.g. dosage of the drug/drug interactions and respective monitoring), “Equipment” (e.g. dialysis) and “Coagulation/Blood Products” (e.g. coagulation problems due to a disorder of hepatic synthesis).

Essential information at a glance in case of emergency

The structure of the card was designed in accordance with the internationally known ABCDE scheme used by most anaesthetists, emergency and intensive care physicians for the prioritised examination and treatment of critically ill or injured patients. The modification of this scheme is intended to represent as best as possible the information on a rare disease as necessary to the acute situation from the perspective of anaesthesiology. The prioritisation of “A” to “E” has deliberately followed anaesthesiological procedures.

The maximum condensed presentation of the most relevant basic information as well as the “Do’s” and “Don’ts” applying to a rare disease is supposed to contribute to patient safety, also in situations with the greatest time pressure (Fig. 2).

The emergency card shall and will not replace the complete recommendations for action – after all, according to its purpose, it also only contains but one fraction of the information contained therein. Yet it is supposed to improve meeting the demands that a rare disease places on anaesthetists in an emergency situation. If needed, time and primarily in advance of elective interventions,

detailed information on the single items can and should still be derived as usual from the respective recommendation for action. The systematic, concise representation of the anaesthesiological risk factors that might be associated with rare diseases are supposed to reduce the likelihood of complications occurring in the treatment of these patients (Fig. 3).

The well-structured presentation of anaesthesiological core subjects intends to draw attention to the risk factors which might be correlated with rare diseases and thus reduce the likelihood of complications during the treatment of affected patients. The chart might also be useful in case of intraoperative complications or a routine re-evaluation. "OrphanAnesthesia" depends on the cooperation of dedicated anaesthetists

and disease experts in order to initiate new recommendations for action, develop the project further with new ideas, thus continually contributing to the anaesthesiological safety of patients with rare diseases. Irrespective of the existing individualised recommendations for action applicable to rare diseases and the availability of an accordingly structured procedure, peculiarities and unforeseen events should always be taken into regard particularly in this special patient clientele [6].

Figure 2

A	AIRWAY / ANAESTHETIC TECHNIQUE	Typical airway difficulties / anomalies / malformations? (Dis)Advantage for general (GA) or regional anaesthesia (RA)?
B	BLOOD PRODUCTS / COAGULATION	Special preparation / storage of blood products necessary? Special haemostaseologic tests / consideration necessary? Coagulation disorders / pathologies?
C	CIRCULATION	Typical cardiopulmonary malformations, pathologies, arrhythmias...? Congenital heart disease / anomaly? Special risk for heart failure? Haemodynamic specifics / risks?
D	DRUGS	Any drugs, that should be avoided? Is any drug dosage necessary? Interactions with typical home medication? Special recommendations for premedication? Risk for MH?
E	EQUIPMENT	Special equipment (or care / assistance) needed for e.g., transport oder within PACU / IMC / ICU?

In the future, the emergency card will provide the attending anaesthetist in condensed form with the most important information on anaesthesiological focus topics. It was structured in accordance with the ABCDE scheme known from emergency medicine and modified in order to better serve the anaesthesiological treatment of emergency patients with rare diseases.

Figure 3

A	AIRWAY / ANAESTHETIC TECHNIQUE	no special airway malformations, but GA may be challenging (due to respiratory status) – availability of tracheal suction (pulmonal bleeding) – anaesthesia only in cases of stable disease if applicable – be aware of severe / refractory hypoxaemic failure due to pulmonal haemorrhage with necessity of VV-ECMO – consider neuraxial / peripheral RA as a feasible alternative if applicable
B	BLOOD PRODUCTS / COAGULATION	be prepared for recurring transfusions in case of persistent intrapulmonary bleeding (sufficient storage of blood products) – be aware of low platelets and fibrinogen in patients undergoing plasmapheresis
C	CIRCULATION	anticipate haemodynamic deviation due to pre-existing hypertension when undergoing anaesthesia – consider IBP (blood gas analysis) and (non-)invasive haemodynamic (to avoid fluid overload)
D	DRUGS	no risk for MH – be aware of secondary insufficiency of the adrenal glands (long-term corticosteroid-therapy) and infectious complications (immunosuppressive therapy) – consider drug dose adaption in case of renal impairment
E	EQUIPMENT	use ultrasound for vessel cannulation / peripheral RA (edema, swollen limbs) – perioperative availability of dialysis may be necessary

An emergency card applying to the Goodpasture Syndrome is shown as an example. In the future, it is supposed to present the most significant information pertaining to the disease to the anaesthesiology team at a glance and in a condensed, prioritised manner. Detailed information may be still be obtained as usual from the recommendations for action. In the future, the emergency card will contain all new and revised recommendations – recommendations already available will be added to the emergency card in the course of time.

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Correspondence address



Dr. med.
Christine Gaik

Klinik für Anästhesie und Intensivtherapie
Universitätsklinikum Marburg
Baldingerstr.

35033 Marburg, Germany

Mail: gaikc@med.uni-marburg.de

ORCID-ID: 0000-0003-1872-6558