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Norwegian Summary Care Record

Critical health information (alert information) in the Summary Care Record

Version 2018

Clinical description and code values

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Notes to the English version

This is a translation of a Norwegian report from the Directorate of Health (from 1.1.2016: Directorate of e-Health) produced as a result of a project for making of a national standard and guidelines for registration of Critical Information /Alert information in the Norwegian Summary Care record (Emergency Care Summary Record). Some of the illustrations and the screen captures from the SCR are still in Norwegian.
The current document is a description of the clinical content and coding system. A separate work has been done to construct a technical description, this is currently in Norwegian but a HL7-FHIR version has been constructed. There has also been some work on construction of archetypes according to this clinical standard.

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Introduction. Background and process

The term “critical information” means information, that in a treatment situation can result in a change to planned intervention, and may save the patient’s life or prevent serious injury. Examples of critical information are drug allergies and other allergies; special disorders such as haemophilia, angioedema, Addison's disease, porphyria, etc.; implants, and previous complications associated with anesthesia, In English language texts, this is often referred to as “alert information”. However, since this is not an established expression in Norway, we have opted to retain the definition “Critical Information”. This is information one particularly wishes to bring to the attention of health care personnel responsible for patient treatment.

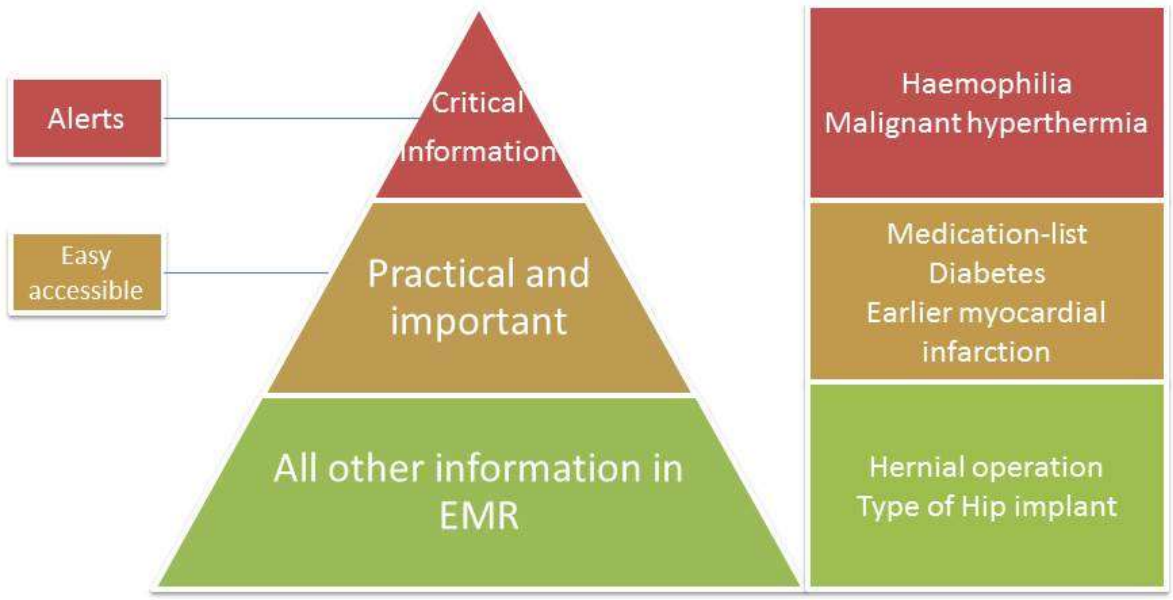

Figure 1 - Illustration of EMR contents

To date, there have been no Norwegian national guidelines or standards for documentation of critical information. Often, there are various professional groups that can edit critical information, and such information is recorded in several locations, including charts, in ongoing medical notes, admission notes and separate modules, which are more or less structured. Providers of electronic patient records /Electronic Medical Records (EMR) have developed their own modules, but the modules lack functionality enabling them to interact with other systems. The terminology used is confusing, and in addition, other health information mixed with what is critical information. It has therefore not been possible to coordinate critical information from different EMRs into the Summary Care Record because of these differences in practice and structure.

With increasing electronic interaction within the health service, and increasing demands for structured documentation, the need to establish a national standard for critical information has become increasingly apparent. In connection with establishment of a national Summary Care

Record, this need for standardization was flagged up by the Government in the whitepaper to the Storting (89L 2011-12).

Whitepaper 89 L (2011–2012)
Changes to the Health Register Act, etc. (creation of a national summary care record etc.)

About critical information: The Department of Health is of the view that critical information must be structured and defined in such a way that it shows clearly which elements of content should be included. It is important to clarify which groups, allergies, diagnoses and implants should be viewed as critical. Code values must be established for critical information such that a selection of the information can easily be transferred to the national summary care record.

As part of the national Summary Care Record project, the Directorate of Health (later Directorate of eHealth) has developed a national standard for critical information. The work commenced in 2012 with the establishment of a three-strong project working party, and an expert group of 17 professionals. The working group performed literature searches, analyses and study visits and created proposals for the structure and content of critical information for the expert group. The expert group had four full-day work meetings where the proposed content was reviewed and amended, and where the group produced further proposals and recommendations. External resource persons were also consulted in separate meetings, to discuss specific problems.

The work of developing a national standard has included assessing existing international models for critical information. The aim was to find a model:

- that can be used without access to a complete EMR
- with a structured, multi-disciplinary content in respect of important and critical information
- that supports grading of the multi-disciplinary information (e.g., patient hypersensitivity reactions) with regard to the degree of severity and the degree of probability
- which, as far as possible, is based on use of international health-science code values
- which can be adapted relatively easily to Norwegian conditions
- and which it was possible to start using in autumn 2013.

There was a natural limit to how many models could be evaluated, but the following were identified and evaluated:

1. KITH-report 42/03 EHR standardization: Cave, Reservations and aims, Practical matters etc.
2. “The Oslo Model” 2010
3. Warning information, (Sweden, 2008)
4. epSOS (2010)
5. Canadian model (2010) (HL7)

Based on the established criteria, we found that the Swedish model for “warning information” established in “NPÖ” (“National Patient Overview”, the Swedish “Summary Care Record”)[1] was the one best suited to meet our requirements. Consequently, this model was used as the basis for the work. Lead author of the 2008 Swedish report (Rikard Lövström) participated in the expert group’s work and helped clarify issues encountered during by the Swedish working-party.

[1] https://sites.google.com/site/warninginformation/resurser-swe/Slutrapport_Varningsinformation_etapp2_080901_ver_1_0.pdf?attredirects=0&d=1

The first version (1.0) of the report concerning the critical information (March 2013) was primarily tailored to the need for a national Summary Care Record, and functionality has been tested in the Trondheim and Stavanger area (August 2013 to autumn 2014). No such work has previously taken place in Norway, and the work has been a combination of preparing technical guidelines (with little in the way of international research), and establishment of an Information- and Communication Technology (ICT) standard. During the test year, there has been a lot of feedback on the entry of "critical information" and constructive suggestions for additions or changes to the code values.

Version 2.0 (autumn 2014) of the report contained a complete updated and technically evaluated code values. Work also took place to specify the category [2]"Change in treatment protocol". This category had not been included in the pilot of the Summary Care Record, but forms part of a complete model, and was implemented in the Summary Care Record in spring 2016.

1.1 National consultative round for "Critical information for the Summary Care Record"

"Critical information for Summary Care Record" (Version 2.0) was submitted for national consultation to key stakeholders in spring 2015 and simultaneously published on Helsedirektoratet.no. The aim of the consultative round was to establish critical information as standard for integration with the Norwegian Summary Care record.
Twenty written submissions were received. One consultative body did not wish to take a position on the content; two supported the report and had no further comments, while 17 stakeholders had a range of comments on its content. However, there were no major principal objections to the work, and almost all stakeholders expressed their support for creation of a standard for how this part of medical information should be recorded. Nor were there any objections to the choice of main model for structuring of the information.

1.2 Clinical challenges in the development of a national standard for critical information

What constitutes critical information for the individual patient varies, depending on the clinical context. What might be considered critical in one situation is not critical in another. At the time of recording critical information, it may be difficult to predict which information will be of critical importance in subsequent situations. This requires that the model for recording information be flexible, and capable of processing many different types of information.

Much of the information will be associated with great uncertainty, e.g. allergies that are not verified by tests or observation. The model needs to be able to cope with such uncertainty, so that uncertain information may also be recorded.

Classification and structured recording of information using codes usually results in more accurate recording than use of free text. It is easier to reuse the data, but a rigid structure may result in compromise when entering information. The accuracy of the information increases with structured data, and a high level of precision is a precondition where critical information is used for automatic warnings (decision-making support) at a later juncture.

Recording data in free text increases flexibility and makes entering data easier for the clinician, but also increases the risk of misinterpretation and ambiguity. It makes automatic warnings and reuse of information for other clinicians more difficult.
Consequently, the model for critical information uses, where possible, structured selection and code values, while free text comments are also possible, giving increased flexibility.

[2] Chapter 4.5.

A system that enables most clinicians responsible for patient treatment to record critical information is considered advantageous. It is also important that the information is of the highest possible quality and validity. The conclusion, therefore, resulted in a main principle: that critical information is recorded only by a doctor. This means the doctor responsible for treating the patient at the time when it would be natural to record the information. In many situations this will be the patient's GP, but it may also be the treating doctor in an acute medical centre or within the specialist health service. No single doctor has overarching editorial responsibility, but the doctor responsible for treating the patient at the material time may instead make changes and additions as necessary.

There is, however, an exception from the general principle that data entries and changes may only be performed by a doctor. In specific individual treatment situations, other health care personnel are delegated responsibility for implementing specific treatment/follow-up. This includes nurses giving outpatient chemotherapy treatments and dialysis, follow-up of heart implant patients, and psychologists who create psychiatric crisis plans for their patients, where they are themselves responsible for the treatment. When other health care personnel are delegated responsibility for—or are responsible for—the treatment, it is important that they can record this treatment and, where applicable, its conclusion. Consequently, the category “Important ongoing treatment/implants” may, in certain situations, be edited by health care personnel other than a doctor.

2 Information categories

2.1 General principles, division into categories

The information model for critical information is based on six major categories. The model is based on the Swedish model “Warning information”, but is adapted to Norwegian conditions. The details of the content within each of the main categories are defined by the Norwegian expert group, and further developed under the pilots of the Summary Care Record in Norway. The contents may differ somewhat from the original Swedish model.

The code values in Summary Care Record are based, as far as is possible, on international classification systems such as ICD-10, ICPC and the ATC-code system. Summary Care Record has also developed a somewhat separate code system and standards for situations where it has not been possible to find suitable existing code values.

2.2 The main categories of critical information:

1. **Hypersensitivity reactions**
   Includes adverse drug reactions and critical allergies. The data recorded includes the type of reaction and its degree of severity, the probable agent, and to what degree the cause(s) of the reaction(s) is/are verified.

2. **Complications of anesthesia**
   Previous complications during anesthesia as a result of a patient condition. Many patients have this information in an "Anesthesia Problem Card”, which they carry with them.

3. **Medical condition**
   Condition/illness that may be difficult to identify in an acute situation, and which may cause serious complications or incorrect treatment if overlooked on investigation and treatment of the patient. Summary Care Record has developed a list of such conditions which should be recorded.

4. **Ongoing treatment and implants**
   Information about ongoing, important course(s) of treatment, which may constitute a risk to health - if not taken into consideration in connection with health assistance given to the patient. Information may be given about the duration, when this is known at the time of recording the information.
   The category also includes critically important implants and grafts.

5. **Changes in treatment routine**
   The responsible healthcare provider and patient may sometimes make decisions that will have implications for the healthcare the patient will receive, which may require deviation from standard procedures and guidelines.
   This includes limiting of life-prolonging treatment, any patient objections to receiving blood products, and agreements about changes in specific treatment protocols.

6. **Infection**
   Includes infectious diseases with consequences for choice of treatment for the patient.

2.3 Additional data (metadata) on recording of critical information

Whenever critical information is entered or changed, the following are recorded:

- Date of entry or change
- Person making the entry (including category of health care personnel)
- Change status
- Expiry date for elements with known expiry date (start and/or stop date).
- The patient's age or date of event, where relevant.
- Source of the information
- Commentary field in free text

2.3.1 Verification of information

It is possible to verify information already entered in the Summary Care Record to ensure that the information is recognized as updated. This will reduce uncertainty associated with the information, when a long time has passed since the entry was made. Similarly, one can confirm that critical information does not exist (negative confirmation), to demonstrate that an empty entry field really should be empty and is not just empty because the entry is incomplete.

2.4 Assessment of 'hypersensitivity reaction' in relation to severity and probability

In the category "Hypersensitivity reaction" the person entering the data must only assess the severity of the reaction that has occurred, not assess what might occur with a subsequent exposure. By describing the reaction at the material time, this gives the next care provider the opportunity to assess the consequences of the information based, among other things, on the patient's needs, the situation, and the risk "there and then". The various disciplines within the health care service have different options in terms of their action in the event of serious incidents. Hence, information about a "less serious reaction" could result in someone in the primary health care service choosing another treatment, while those working on an intensive care unit will have the preparedness and opportunity to be able to manage the consequences of a possible hypersensitivity reaction.

The person entering the data must also decide how certain one may be about there being a probable connection between the stated cause and the reaction.

2.4.1 Grading of severity

Only hypersensitivity reactions and allergies that may be relevant in a medical acute situation should be recorded, and the entry must be graded. Grading means that the data recorder must state the severity of reaction that has taken place.

Severity:

a) Severe reaction
   - Reaction that was potentially life-threatening or has caused injury to health with a given duration.

b) Less severe reaction
   - Reaction that was troublesome or unpleasant, but posed no risk to life or injury to health

A description of the actual event is what provides the greatest security because it is impossible to predict whether a "less serious reaction" may be life-threatening on the next exposure.

2.4.2 Probability

There must be thought to be a connection between the described cause and the patient's reaction. How probable this connection is thought to be, will be graded.

Grading of probability (certainty):

1. *Suspected*
   All connections that cannot be excluded, including when clinical observation is not uniform
2. *Probable*
   The connection is more probable than other possibilities, but documentation, as described for "confirmed" is lacking
3. *Confirmed*
   The connection is documented by analyses, including provocation test, investigation by allergologist or similar
4. *Disproven*

Reactions occurring under the influence of several agents should be recorded as "Suspected" in all cases where it is not overwhelmingly probable or known that the reaction is due to *one* specific agent.

The degree of probability should be changed, as knowledge of the patient's situation changes. A change to "Disproved" status means that an attempt has been made to confirm the hypersensitivity reaction through laboratory tests or provocation tests, but that these were negative, and that the patient most probably is not suffering from this.

2.4.2.1 Why show "Disproved" information?

When a suspected hypersensitivity is disproven, it no longer need be notified, but the information nonetheless remains part of "critical information" because "negative confirmation" in this context is medically important information.
Suspected, severe hypersensitivity recorded in the Summary Care Record may remain active in one or more electronic medical records after it has been recorded as disproven in the Summary Care Record. *Currently, there is no automatic exchange of data between the local medical records and Summary Care Record.* When the patient attends for treatment and there is an entry in the local medical record relating to a suspected serious allergy, and where there is no record of allergy in the Summary Care Record, it is important that the suspicion be shown as "disproven", in order that it can be re-entered in the Summary Care Record. Consequently, the next care provider should be able to see that an entry has been made in Summary Care Record that was initially entered as "suspected" but later disproven.
Disaffirmation of a suspected allergy may also mean that the reaction that resulted in the suspicion is caused by another allergy, as yet unknown. This should lead to increased caution. An example of this is a patient who has a serious reaction under general anesthesia. The suspected allergy is entered in the anesthetic notes, but this suspicion is subsequently disproven. It is then important to be clear that the patient has reaction to something ELSE, which at the time is unknown, and extra precautions should be taken in any subsequent anesthesia.

2.5 Source of information

As a general principle, the source of any information should be stated, where possible. Alternative source statements:

1. *Information from the patient*
   Information from the patient, without the treating physician having been able to verify this objectively by tests or reliable documentation.
2. *Information from relatives*
   Information from relatives, where the patient cannot themselves verify this. Applies in particular to minors, and patients who are unable to self-report.
3. *Taken from existing notes*
   Information recorded in the EMR or other documents, without one knowing with certainty who the original source of information is, or where the person who entered the information cannot be contacted where necessary to confirm the information.
4. *Stated by responsible care provider*
   The responsible care provider/health care personnel (doctor, psychologist, nurse etc.) can confirm that the information is correct. Often used when recording in categories other than hypersensitivity reactions, where one cannot say that the information is “observed” (critical diagnoses, treatments etc.). May also be used when the person entering the information has not personally observed a reaction, but is notified of it by another responsible health care worker with first-hand knowledge, e.g. via the discharge notes.
5. *Observed by treating doctor*
   Used primarily in the “hypersensitivity reactions” category, when the person entering the information about a reaction has also observed it.
6. *Result of tests/analyses*
   Used when a reaction is confirmed by analysis or tests (e.g. provocation testing). May also be used where there is an investigation and conclusion from an allergologist.
7. *Other*
   Other sources of information that do not naturally fit into one of the above code values. Comment with explanation should be given.

3 Encoding and description of the individual information elements

In this chapter, each category of information is described in detail. The chapter will provide sufficient information to enable the complete code values to be entered, and provide sufficient information about the background for the choices made in the development of critical information as a shared national standard.

3.1 Hypersensitivity reactions

3.1.1 Drug reactions

Knowledge of prior serious reactions to drug treatment can be crucial when choosing further treatment for the patient and this is important information, which can be passed on via the Summary Care Record.
Information passed on in the Summary Care Record shall provide descriptions that are as neutral and factual as possible. It is important that the Summary Care Record does not interpret, or make assessments about the information entered therein, since the assessment will depend on the actual situation. The tradition in Norway in the past decade has been to use the term “CAVE». «Cave" is the imperative form of the Latin verb "CAVEO" and means "beware of" or "beware»[3]. In practice, this is interpreted as a caution that the treatment is contraindicated. In the Summary Care Record, the decision has been made not to use the term “CAVE”. It is important that the next care provider knows which drug reaction the patient experienced, but whether or not information of a prior hypersensitivity reaction will mean that this drug will not be used in the next treatment situation will vary depending on the situation and the indication. Not using CAVE as a term is a conscious choice to distinguish the new categorization from the old, and will require some extra training.
The Summary Care Record will describe what actually happened, i.e. the suspected agent, the reaction the patient experienced, the severity, and how certain this information is. The Summary Care Record will not give guidelines for what consequences this will have for subsequent treatment, since this will have to be determined by the responsible care provider in each individual situation. We will, however, accept that the Summary Care Record may contain information about how a problem has been solved, or advice and experience of possible alternative treatments. We know that this type of information is highly significant in terms of whether one changes planned treatment.

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3.1.1.1 Coding and recording of drug reactions

The Summary Care Record primarily facilitates recording of the name of the medicinal product, when this is known, structured by searching in FEST.[4] The Summary Care Record stores the name of the medicinal product in the form of “Brand-ID” and may subsequently present this, together with the ATC code and active substances where relevant, to the EMR for decision-making support.
Where the name of the medicinal product is not known, one may only enter the relevant ATC-code, possibly at parent level, which gives the option of structured entry of unclear information.
It is possible to record the *active substance* in a structured search where there are different ATC-groups with the same active substance. It is also possible to indicate that only the entered medicinal

[3] Myren et al; Watchdog in the journal, Tidsskr Nor Legeforen 2014; 134: 1486-7
[4] “Shared expedition- and decision-making support”. Database published and maintained by the Norwegian Medicines Agency.

product shall be flagged. This is done in those situations where it is felt that the patient reacts only to the excipients of a given preparation.

The following coding may, therefore, be used to state the drug to which the patient has reacted:

> ***Example of recording of uncertain information****: If we know that the patient reacted to a contrast agent given during MRI, without knowing what was used, one may record hypersensitivity to ATC-code V08C "Contrast media for MRI" until one has obtained more detailed information about which contrast agent was used.*

1. Brand name (Brand-ID after structured search in FEST)
2. ATC code (7, 5 or 4 digits/spaces)[5]
3. Active substance-ID (substance codes from FEST)
4. Excipients (logical variable, must be combined with (1)

Rules:

1. Brand names may be recorded alone, but will at the same time automatically result in storage of the 7-digit ATC-code
2. The ATC-code can be entered to 7, 5 or 4-places. Must be entered alone.
3. The active substance is stored after searching in FEST. Must be entered alone. Other codes are not stored.
4. Excipient reaction - logical variable, must be inserted together with a brand name (Brand-ID after structured search in FEST). Does *not* result in storage of ATC or active substance-ID.

3.1.1.2 Decision-making support

The Summary Care Record will enable giving of the following information to an EMR:

1. Brand-ID from FEST:
    a. Alone, but then paired with logical variable "Excipient-reaction»
    b. Together with 7-digit ATC code and active substances

2. ATC-code
    a. Alone (7, 5 or 4 digits)
    b. 7 digits, together with Brand-ID

3. Active substance-ID
    a. Alone

Examples of logic in EMR for warning of hypersensitivity reactions on new prescription:

1. If ATC-code is set: warn on all encounters with same ATC (7, 5 or 4 digits)
2. If substance-ID is set: warn on all encounters with same substance-ID
3. If Brand-ID *and* Excipient-reaction are set: Warn of same Brand-ID only

[5] Some ATC-codes can be registered with only 3 digits/spaces.

3.1.2 Other reactions/allergies

Recording of allergies and other should include reactions that may be critical in the context of a medical acute situation. The entry will include the agent, the type of reaction and degree of severity, and to what degree the cause(s) of the reaction(s) have been verified.

The aim of Summary Care Record is to be able to provide a structured choice when entering critical information. For recording of allergies and other reactions, significant mapping work has taken place to ensure that the structured choices are as extensive as necessary, but at the same time remain within what is clinically beneficial and practically functional.
Several databases that include described allergens are considered, including the English database "The Allergen Database"[6] which proves too detailed.
The Norwegian Food Allergy Registry shows that there are a few allergens that account for more than 95% of all serious reactions. On this basis, we have developed our own "shortlist, using these allergens as point of departure. It is possible to record comments in free-text to ensure flexibility in terms of entry options. In addition to reaction type are recorded severity and degree of probability, to ensure that the information is correct, in the same way as for drug reactions.
The recommended guidelines for recording entries are:

Here, only allergies that may have serious consequences in a treatment situation if the allergy is unknown, are recorded. Only serious allergies relevant in a medical acute should be recorded.

Shortlist of important allergies:

- Milk
- Eggs
- Wheat
- Shellfish
- Fish
- Peas
- Soya
- Lupine
- Fenugreek seed
- Peanuts
- Nuts other than peanuts
- Poppy seed
- Insect toxin
- Latex
- Other important allergies (specified in free text)

3.1.3 Reaction types

The international (EEACI and WAO) definition of hypersensitivity[7] is: "Objectively reproducible symptoms or signs, initiated by exposure to a defined stimulus that is tolerated by normal subjects". It is important to record in structured form, which type of reaction the patient had to the allergen that was thought to be responsible for the reaction. Such an entry will make possible a standard stated severity for the event, which is more uniform than if it were entered in free-text.

The definition of hypersensitivity includes both allergic reactions and type-B-adverse effects, i.e. side effects due to the medicinal product's unknown pharmacological effects. The types of reaction that can be recorded must, therefore, include both allergic reactions and the most important type-B-side effects. Recording of hypersensitivity reactions to drugs shows that skin reactions dominate[8]. The list of reaction types must, therefore, include more detail in respect of skin reactions than other organ systems.

[6] Website: allergen.csl.gov.uk
[7] http://www.worldallergy.org/professional/allergic_diseases_center/nomenclature/english.php
[8] Presentation by Bernstein and Greibe, e-sundhedsobservatoriet 2012, DK

Coding system for reaction types:
(See description in table 1, page 17)

- **Anaphylaxis**
  - Anaphylactic reaction = Suddenly occurring reaction affecting at least 2 organ systems
- **Reaction involving the cardiovascular system**
  - Hypotension
  - Serious arrhythmia
- **Reaction affecting the apparatus of breathing**
  - Laryngeal edema
  - Asthma
  - Unspecified difficulty of breathing
- **Central nervous system reaction**
  - Seriously affected level of consciousness/confusion etc.
  - Generalized seizures
- **Reaction affecting skin/mucosa**
  - Angioedema/severe generalized urticaria
  - Other severe skin reactions such as Stevens-Johnson syndrome, epidermolysis, severe bullous reactions, vasculitis etc.
  - Less severe reactions such as itching/swelling/local urticaria/erythema
  - Irritation of the eyes, nose, throat
- **Reactions affecting the GI-tract**
  - Vomiting/diarrhea/abdominal pain
- **Other reactions**
  - Liver failure/hepatic impairment
  - Renal failure/renal impairment
  - Blood: aplasias/dysplasias/cytopenias
  - Rhabdomyolysis
  - Other severe reaction
  - Other less severe reaction

3.1.3.1 Unknown reaction

During piloting of the Summary Care Record, a need arose to be able to record preliminary and somewhat deficient historical information about a suspected hypersensitivity reaction. This is because it should be considered in the event of a new prescription. There are situations where one, at the time of recording the data, cannot state the type of reaction, because this is historical data that it is not possible to relocate at the time. This may be medical record entries such as “CAUTION: penicillin”, or information from relatives that the patient “can’t tolerate” a specific drug; in both instances without being able to state specifically which reaction the patient has had. This means, therefore, that one can state “Unknown reaction”, but one must then state the probable degree of severity and be prepared to write a comment in order to aid the next provider’s assessment. Use of "unknown response" should be limited and used only as a preliminary entry until more information is obtained.

3.1.3.2 Reaction from more than one organ system

If the patient has had more than one reaction to a hypersensitivity event, this should be recorded as "anaphylactic reaction" if there is "sudden reaction from at least 2 organ systems". Examples of this may be asthma and blood pressure drop. If there are 2 reactions that cannot be seen in a context and "suddenly occurred" eg. drug reaction with both affected liver and kidney function, one should register the most significant reaction, (the most severe) and in the comment field describe what else happened.

3.1.4 The relationship between reaction types and degree of severity

Grading of severity is necessary in order to be able to give a correct description of the reaction that has taken place. The degree of severity is recorded based on which type of reaction the patient has had, and is flagged as either *Critical* or *Important*. When choosing a reaction type, a standard degree of severity is entered, depending on the type of reaction, but this can be overridden by the physician making the entry in special instances. A *Less severe* reaction may—on the next exposure—result in a critical or life-threatening reaction, and the grading is, therefore, only a description of what has taken place, and not of how this should be managed on the next exposure.

The code values of reaction types is constructed in such a way that one DOES NOT usually have to state the degree of severity in addition to the reaction type. Consequently, some reaction types are stated as “severe” or “less severe”. The user cannot change the degree of severity for these reaction types, so another reaction type must be chosen.

Table 1 Degree of severity - indicative table (S = Severe LS = Less severe)

Name	Degree of severity	Description
Anaphylaxis		
Anaphylactic reaction	S	Sudden onset reaction affecting 2 or more organ systems shall be entered as anaphylaxis
Reaction involving the circulatory system		
Hypotension	S	Sudden drop in blood pressure that cannot be predicted on the basis of known pharmacological properties
Serious arrhythmia	S	Includes serious ventricular arrhythmias, cardiac arrest, serious AV-block and prolonged QT-interval
Reaction involving the respiratory system		
Laryngeal edema	S	Includes all acute swelling of the upper airway with obstruction
Asthma	S	Obstruction of the lower respiratory tract
Unspecified difficulty breathing	S	Other stridor/dyspnea, which cannot be classified as laryngeal edema or asthma (see above)
Central nervous system reaction		
Effects on level of consciousness/confusion	S	Includes unconsciousness. The reaction must have occurred on a dose of the medicinal product that does not normally cause such a reaction
Generalized seizures	S	Includes all serious extrapyramidal reactions. Seizures thought to be functional should not be entered
Reaction affecting skin/mucosa		
Angioedema/severe urticaria	S	Swelling of the face/lips/eyelids and/or generalized severe urticaria. Limited urticaria with no general effects are recorded as "less severe skin reaction"
Other severe skin reactions	S	Includes, among other things, Stevens-Johnson syndrome, epidermolysis, vasculitis, severe bullous skin diseases etc.

Less severe skin reaction	LS	Includes itching, local swelling, limited urticaria and erythema without constitutional symptoms
Irritation of the eyes, nose, throat	LS	Applies only to irritating reactions. Reactions causing obvious breathing difficulties are recorded “reaction affecting the respiratory system”
Reactions affecting the GI-tract		
Vomiting/diarrhea/abdominal pain	LS	Any gastrointestinal reactions, which cannot be expected given the medicinal product’s known pharmacological properties
Other reactions		
Liver failure/hepatic impairment	S	Any gastrointestinal reactions which cannot be expected given the medicinal product’s known pharmacological properties
Renal failure/renal impairment	S	Compromised renal function, which cannot be anticipated based on the medicinal product’s known pharmacological properties at a normal dose.
Blood: Aplasias/dysplasias/cytopenias	S	All serious changes in the blood picture, which cannot be anticipated based on the medicinal product’s known pharmacological properties at a normal dose.
Rhabdomyolysis	S	Rapid breakdown of skeletal musculature, which cannot be anticipated based on the medicinal product’s known pharmacological properties at a normal dose
Other severe reaction	S	Other reaction that has been life-threatening or has caused serious damage to health. Remember to specify in free text
Other less severe reaction	LS	Another reaction that was distressing for the patient, but caused no serious harm to health. Remember to specify in free text

Table 1 Degree of severity - indicative table (S = Severe LS = Less severe)

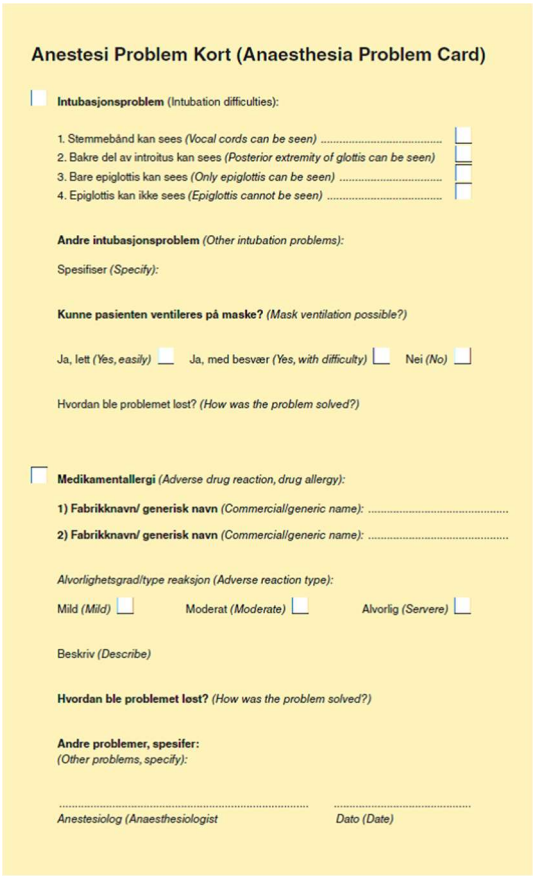
3.2 Complications of anesthesia and other previous treatment complications that are conditional in relation to the patient

Giving anesthesia is a risk-laden procedure, which of itself has no medical benefit. Consequently, the risk should be the lowest possible.[9]In the event of known, previous complications under anesthetic as a result of a patient-related factor, the risk may be reduced by taking precautions in the form of alternative equipment, a change in procedure or increased staffing, adapted to the anticipated complications. The anticipated complications can be perceived on the basis of information about known problems and symptoms, but also whether the patient previously experienced complications during anesthesia.

Internationally, having an anesthetic problem card, which sums up these most important complications, is a means of communicating this problem.[10]The card is given to the patient, who is asked to carry it on their person and to show it to the doctor prior to planned interventions. Summary Care Record makes information on the card available electronically.

Anaphylaxis during anesthesia is a separate section on the anesthetic problem card. The most important allergens are peripherally acting neuromuscular blockers, latex, antibiotics and chlorhexidine. This should be documented in the information category “Hypersensitivity reaction”.[11]

[9] Botney R. Improving patient safety in anesthesia: a success story? Int J Radiat Oncol Biol Phys 2008; 71: P.182–6.
[10] Hjemmeside: http://www.nafweb.no/index.php?catid=7:ny-pa-nafweb&id=80:problemkort&option=com_content&view=article
[11] Anafylaksi under anestesi A B Guttormsen et al Tidsskr Nor Legeforen 2010; 130:503-6



Anestesi Problem Kort (Anaesthesia Problem Card)

Intubasjonsproblem (Intubation difficulties):

1. Stemmebånd kan sees *(Vocal cords can be seen)* .....
2. Bakre del av introitus kan sees *(Posterior extremity of glottis can be seen)*
3. Bare epiglottis kan sees *(Only epiglottis can be seen)* .....
4. Epiglottis kan ikke sees *(Epiglottis cannot be seen)* .....

Andre intubasjonsproblem *(Other intubation problems):*

Spesifiser *(Specify):*

Kunne pasienten ventileres på maske? *(Mask ventilation possible?)*

Ja, lett *(Yes, easily)* Ja, med besvær *(Yes, with difficulty)* Nei *(No)*

Hvordan ble problemet løst? *(How was the problem solved?)*

Medikamentallergi *(Adverse drug reaction, drug allergy):*

1) Fabrikknavn/ generisk navn *(Commercial/generic name):* .....

2) Fabrikknavn/ generisk navn *(Commercial/generic name):* .....

Alvorlighetsgrad/type reaksjon (Adverse reaction type):

Mild *(Mild)* Moderat *(Moderate)* Alvorlig *(Servere)*

Beskriv *(Describe)*

Hvordan ble problemet løst? *(How was the problem solved?)*

Andre problemer, spesifer:
(Other problems, specify):

....

Anestesiolog (Anaesthesiologist *Dato (Date)*

Figure 2: The Norwegian anesthetic problem card, ref. Association of Norwegian Anesthesiologists

Consequently, Summary Care Record will facilitate recording of:

- **Intubation problems**
  - Grading of anatomical access for intubation (Cormack & Lehane)
    1. Grade 1: Vocal cords visible
    2. Grade 2: Posterior extremity of glottis seen
    3. Grade 3: Only epiglottis visible
    4. Grade 4: Epiglottis not visible
    5. Other intubation problems
       - Specify in free text
  - Able to mask ventilate?
    1. Yes, easily
    2. Yes, with difficulty
    3. No
    4. Not stated
  - How the problem was solved
    - Specify
- *(Drug allergy - Display and enter in structural form under Hypersensitivity reaction)*
- *(Medical condition - Display and enter in structural form under Hypersensitivity reaction)*
- **Other anesthetic problems**
  - Specify in free text

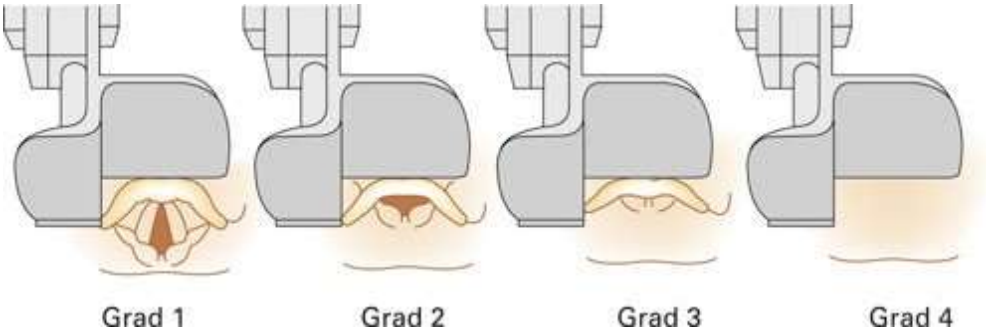


Figure 3 Classification of views of the larynx obtained by direct laryngoscopy by Cormack & Lehane[12,13]

3.3 Medical condition

Medical condition in this context means a *condition that may be difficult to identify in an acute situation, and may cause serious complications or mistreatment if overlooked.*
When evaluating a patient in an acute situation, the entire patient history is of importance. However, some conditions and diagnoses are more significant than others because they do not normally form part of the normal assessment of possible diagnoses. Whether a patient has diabetes or suffered myocardial infarction several years ago is of course important; however if one is not actively warned of this, it is unlikely to harm the patient because these possibilities are taken into account as part of the diagnostic process. But if a patient with chronic adrenal insufficiency is admitted in Addisonian crisis associated with another acute event, the chances are significant that the adrenal insufficiency will not be identified if the investigating health care professional is not notified that the patient has this condition. A number of such conditions exist, where there is significant risk of overlooking them, because they are not normally part of the diagnostic assessment.
These conditions have been difficult to define in more detail. A search of the literature has been unable to provide lists of conditions that meet these criteria, and nor have Summary Care Record projects in our neighboring countries managed to create lists of those conditions, which should form part of the information recorded.
(The closest we come to a classification is the “ASA”, which is used to document anesthetic review before surgery, among other things. In 1941, the authors faced the same challenge[14]. The aim was to classify physical conditions, with a view to creating a uniform system for statistical analysis. In practice this is in use today to state anesthetic risk, even though a number of other factors will influence this assessment, such as the type of procedure, surgical variation etc. In addition, the ASA-classification also changes with acute illness, such that an ASA-classification is a “snapshot in time”.)

Some disorders may also have a natural range - from insignificant or episodically irritating symptoms, to life-threatening illness - one typical example being asthma.
Consequently, Summary Care Record is designed such that all conditions can be entered, but only a selection of conditions is notified as “critical”. Other conditions are notified as “important”.

3.3.1 Detailing of critical diagnoses - “The absolute list”

There are a number of conditions, usually rare, that meet the criteria for the definition of a critical medical condition, as described previously. There are also a number of conditions that, for some patients, should perhaps be entered, and finally, some that should only be recorded by exception. There is no complete list of conditions that meet the criteria for a critical medical condition, and

[12] Cormack RS, Lehane J. Difficult tracheal intubation in obstetrics. Anaesthesia 1984; 39: 1105–11.
[13] Hvordan oppnå fri luftvei? Bjerkelund et al Tidsskr Nor Legeforen 2010; 130: 507-10
[14] Saklad M. Grading of patients for surgical procedures. Anesthesiology 1941; 2:281-4

Summary Care Record has, therefore, chosen a solution whereby the users can enter conditions they feel meet the criteria.
However, an “absolute list” has been established, which is a list of conditions it is felt meet the criteria for critical information in the majority of situations. As starting point, the list was established based on proposals from a number of clinical specialists, and updated during the pilot period and at the national consultative round for critical information in spring 2015. It is anticipated that the list must be adapted to needs over time by diagnoses being added or removed.

The movement of entries is illustrated in figure 4.

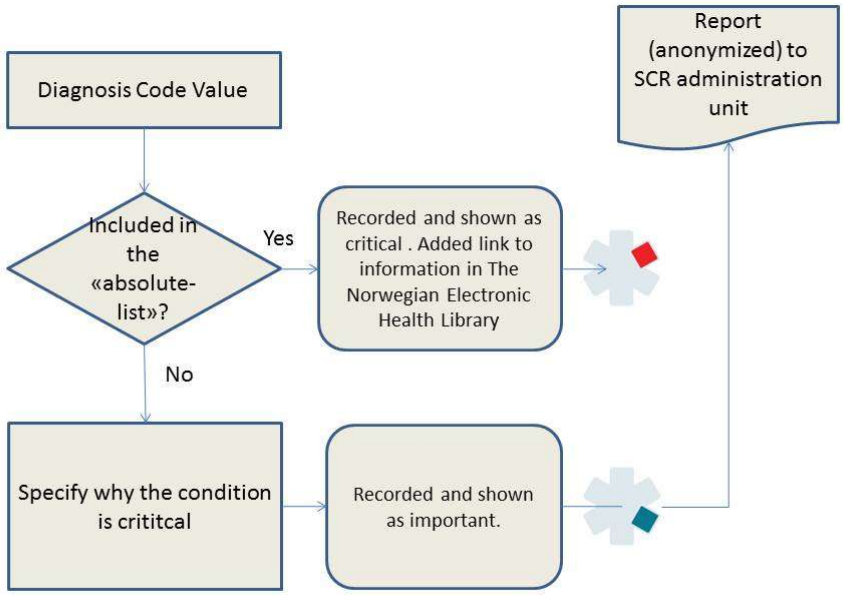


Figure 4. Logistics on recording of critical diagnosis

3.3.1.1 Recording of critical diagnosis

It is possible to record a medical diagnosis/condition as “critical diagnosis” in a field for searching diagnoses. The search takes place in “the absolute list” and in the ICD-10 diagnosis code values, and including synonyms, common Norwegian disease names and selected codes from the ICPC 2 code values, which is used in the primary health service.
Where the recorded diagnosis/condition belongs to “the absolute list”, i.e. list of previously defined conditions that meet the criteria, the condition is entered and notified as critical in the Summary Care Record and there is added a link to more information in The Norwegian Electronic Health Library

If the condition does not belong to this list, the user is asked to explain why the condition is critical by choosing from the list of consequences caused by the condition, see the list below. The condition is recorded and notified as important in Emergency Care Summary Record. If the doctor *cannot* specify at least one choice from the list he has the option of justifying the entry in a free text field.

In those cases where the condition is not on “the absolute list”, notification of the entry is sent to the Emergency Care Summary Record’s administration unit. Here, the relevant entries are stored in anonymized form, and a group of medical professionals assesses whether the conditions in question should be included in “the absolute list”.

The condition must be difficult to recognize, and should also satisfy at least one of the following criteria:

1. Can affect the level of consciousness
2. Can have serious effects on respiration
3. Can result in a bleeding tendency
4. Can cause circulatory failure
5. Risk of problems during anesthesia
6. Risk of complications during surgery
7. Risk of life-threatening complications if drug-treatment changes

Table 2 - list of consequences of the condition

In this way, a list is gradually built up of conditions, which the clinicians feel should be considered "critical" as per the definition. This makes it possible to adapt the list based on actual clinical needs and not only based on theoretical assessments. It is important that this type of warning always be developed in step with knowledge and practice.

3.3.1.2 "The absolute list" - list of critical conditions that should be entered in Emergency Care Summary Record

"The absolute list" is the result of input from a number of specialists, piloting of Summary Care Record for two years, and significant useful input associated with national consultation. This will be continuously updated based on experience and need, in step with further use of Summary Care Record and medical developments. Consequently, the list is dynamic and under continual development.

The absolute list, version dated September 2015, is included in Enclosure B

The list consists of

- *the disease nomenclature ("term")* which is the normal Norwegian or international name (when no Norwegian name exists, or is insufficient)
- *synonymous key words* which are other terms used about the condition, including, to a degree, "colloquial" terms. May incorporate ICPC codes in cases where these are present.
- *ICD-10 codes* is/are the ICD-10 code(s) describing the condition.
- *Rationale* is a brief description of why the condition is deemed "critical". Provides a description and brief advice about management of patients in an acute situation.

Term	**Norwegian synonyms/key words**	**Matching ICD-10 codes**	**Rationale**
Angioedema	Non histamine-induced angioedema Angio-neurotic edema Non-histamine angioedema Quincke's edema Hereditary angioedema HEA	T78.3 Angioneurotic edema	The patient may develop acute swelling of the dermis and mucosa of the face and upper respiratory tract. Can have a turbulent course, and respiratory failure may result if treatment is not commenced. Can be difficult to intubate.

Example of condition in "the absolute list"

3.3.1.2.1 ICD-10-codes in the "absolut list"

ICD-10 codes can partly be entered as keywords and partly as "matching ICD-10 code". ICD-10 codes that uniquely define the state are listed as "matching ICD-10 code". In such a case, the SCR will perceive that the condition in the absolute list is selected regardless of whether you enter the ICD-10 code or make the selection in the absolute list. ICD-10 codes that are only specified as keywords do not define the state unambiguously and therefore cannot be set equal to the term in the absolute list, but will only provide hits in the list during the registration process.
Ex: The *"Muscular dystrophy / myopathy"* absolute list entry has "*G71.0 Muscle Dystrophy*" as "matching ICD-10 code, while the ICD-10 code "*G71.9 Unspecified Primary Muscle Disease*" is only a keyword.

3.4 Ongoing treatment and implants

This category is divided into two sub-groups “ongoing important treatment” and “Implants”. Recorded here is information about ongoing important courses of treatment and implants, which may constitute a risk to health if not considered in connection with health assistance given to the patient.

3.4.1 Ongoing important treatment course

Knowledge about current courses of treatment may affect choice of treatment and other health interventions, it can contribute to the right diagnosis, and there may be information about treatment that should not be discontinued. Courses of treatment that are not shown in the patient’s list of medication are particularly worthy of mention here.

Treatments that can be recorded include:

1. Chemotherapy
2. Immunosuppressant/-modulating treatment
3. Radiotherapy
4. Important anticoagulant therapy
5. Dialysis
6. DAR (Drug-assisted rehabilitation)
7. Participation in clinical pharmacological trials
8. Established psychiatric care plan
9. Ongoing outpatient treatment - pain clinic
10. Ongoing outpatient treatment - Chelate treatment
11. Ongoing investigation/follow-up of suspected or verified malignant disease
12. Other outpatient treatment
13. On list for transplantation
14. Other important treatment

The list is not exhaustive, and may be expanded as necessary. (Some of the items on the list are not strictly speaking "ongoing treatment" but are included because they are important to know, and do not appear elsewhere in the patient's Summary Care Record). The principle is that it should only include recorded treatment that does not appear on the drug list, or which is so critical that it must be specially marked, either due to risk of erroneous discontinuation/change or because attention must be drawn to the treatment in case the drug list is incomplete.

3.4.2 Implants

This category also includes transplants and permanent prostheses and foreign bodies. The reason for the entry is to provide information, which in an acute situation could influence investigation- and treatment options. It is important to limit entries to those implants/prostheses that are meaningful in

this context, i.e. that are significant in terms of diagnosis and choice of treatment in an acute situation. The level of detail in the code values is adapted for this purpose.

3.4.2.1 Classification of implants etc. in national Summary Care Record

Head - neck - CNS

1. Ear implant (including cochlear implant)
2. Metal in the eye (foreign body, etc.)
3. Other implant in structures adjacent to the eye
4. Intracranial vascular clips, etc.
5. Shunt etc. in the central nervous system
6. Neurostimulator
7. Implant in the mouth, nose, throat
8. Tracheostomy

Cardiovascular

9. Pacemaker
10. Implanted defibrillator (ICD)
11. Mechanical heart valve
12. Other implanted heart valve
13. Coronary stent
14. Stent in other than coronary vessels
15. Vascular prosthesis

Abdomen

16. Stent, catheter or implant in the upper urinary tract
17. Biliary stent
18. Stent in other hollow organ
19. PEG tube

Pumps

20. Medical pump
    (Electronic medical pump or other injection/infusion equipment)

Orthopedic

21. Orthopedic prosthesis or implant
    (Only prosthesis or implant significant in clinical acute context)

Transplants

22. Transplanted kidney
23. Transplanted kidney
24. Transplanted lung
25. Transplanted heart and lungs
26. Transplanted liver
27. Transplanted bone marrow, pancreas, intestine, stem cells
28. Other transplanted organ or tissue

Other

29. Other implant
    (Other implant, foreign object, prosthetic or aid worn on the body)

3.4.2.2 About recording of implants, grafts, permanent prostheses and foreign bodies

This is an extensive area and there are many patients who have implants or dentures that appear in the above code values. It is important to have the underlying thought behind “critical information” in one’s head when determining whether a patient’s implant should be recorded. *The aim of the*

recording critical information is to increase patient safety. An implant of significance only in terms of medical history should not be entered in Emergency Care Summary Record.
This can be illustrated with some examples:

- Orthopedic prosthesis/implant
  Hip- and knee prostheses do NOT normally need to be entered. This has little value in an acute medical situation, and the large number of patients will reduce the warning value of critical info.
  Rod fixation of the spine, however, is an orthopedic "implant", which should be entered since it might affect the method of maintaining a clear airway and assessment of "stiff neck".
- Coronary stent
  These are of greatest significance when the warning is entered in the period immediately following implantation, while the risk of restenosis remains high. When the stent is endothelialised, it is of value only in terms of medical history (relates to coronary artery disease). These should only be notified for 9-12 months after insertion.

3.5 Changes in treatment protocol

The health service has many fixed procedures for treatment and investigation. In some situations one should be aware that good reasons exist for deviating from the fixed procedures. The responsible health care personnel and the patient may, in some cases, make decision that will be significant for which health care support the patient will have, and may require deviation from normal procedures and guidelines. This category can be used to record which important changes in normal treatment routine are agreed, and Summary Care Record can, in some situations, notify of this type of procedure changes.

Two categories are defined for change in treatment routine in the Summary Care Record:

1. Limitation of life-prolonging treatment
2. Other changes in procedure

3.5.1 Limitation of life-prolonging treatment

3.5.1.1 Legislation

A dying patient, in certain situations, has the right to refuse life-prolonging treatment. The right means that the patient can refuse to receive treatment whereby there is no likelihood of recovery or cure, but only a degree of prolongation of life, which in reality is a prolonging of the ongoing process of dying.

This appears in the Norwegian Act on Patient- and User Rights, § 4-9.

Life-prolonging treatment in this context means all treatment and all measures that may delay a patient's death. Examples of this include cardiopulmonary resuscitation, other respiratory support and inotropic drugs, nutritional- and fluid therapy (intravenous or via nasogastric- or gastrostomy tube (PEG), dialysis, antibiotics and chemotherapy.[15]
.

[15] IS-2091, chapter 1, http://helsedirektoratet.no/publikasjoner/beslutningsprosesser-ved-begrensning-av-livsforlengende-behandling/Publikasjoner/IS-2091.pdf

3.5.1.2 Decision-making process on limitation of life-prolonging treatment

In the Norwegian Directorate of Health's guidelines: "Decision-making processes in limitation of life-prolonging treatment", IS-2091[16]the decision-making process for not giving full life-prolonging treatment is described in detail. The physician responsible for treatment at the material time is the person responsible for deciding whether to refrain from providing treatment. The guidelines refer to the Act on Patient- and User Rights, which gives an informed- and legally competent dying patient, who wishes to limit possible life-prolonging treatment, the right to have their wishes respected.

If an adult patient is legally incompetent to give consent, the doctor's decision shall be based on what is medically-determined to be in the patient's best interests, and on what one assumes the patient's own wishes would have been. An assessment of what the patient's possible wishes might be may be based on:
a) information from next of kin and healthcare professionals who know the patient well
b) written treatment wishes and
c) consultation with other qualified personnel.[17]

The physician-in-charge of the patient shall make a concrete assessment of whether any recorded treatment request applies to the situation in question, or whether the patient may have changed their mind. The written statement shall constitute one of several sources of information when a decision needs to be made about reducing or withdrawing treatment.

Health care personnel who have to decide whether to initiate life-prolonging treatment must be sure that the patient was of sound mind and understood the content of the offer of treatment.

When a decision is made to cease life-prolonging treatment, the treatment shall focus on the patient's symptoms and care needs, and the need to provide existential and psychological support. The medical intervention that shall be commenced or concluded may include a reduction in medication, withholding antimicrobial treatment and nutritional/fluid therapy, abstaining from technical organ support and further investigations and diagnosis.

3.5.1.3 Entering the right to oppose life-prolonging treatment in the medical record.

If a patient wishes to oppose life-prolonging treatment based on the Act on Patient- and User Rights, §4-9, this shall be recorded in the patient's medical record[18] (EMR). By entering the wish for treatment in the notes in consultation with health care personnel, e.g. a GP, one ensures that the patient receives the necessary information about the meaning of the declaration.

In order for information to be available to other health care personnel who need the information, the patient's wish for treatment should also be recorded in the patient's Summary Care Record.
In the Summary Care Record, such a wish for treatment should appear under "Change in normal treatment protocol".

Where the patient has stated specific wishes or, where other concrete assessments have been performed, these should be entered as free-text comments.

The Norwegian Directorate of Health suggests the following standard text in Summary Care Record:

[16] IS-2091, http://helsedirektoratet.no/publikasjoner/beslutningsprosesser-ved-begrensning-av-livsforlengende-behandling/Publikasjoner/IS-2091.pdf
[17] IS-2091 Section 7.2.3, "The adult patient who is legally incompetent"
[18] Regulation relating to patient records, § 8 paragraph j.

"I do not want life-prolonging treatment if I am dying, in other words, am in an ongoing process of dying and will die in a short time, and I am unable even to convey a wish for treatment. The situation includes both acute conditions/events and planned/foreseeable conditions/events."

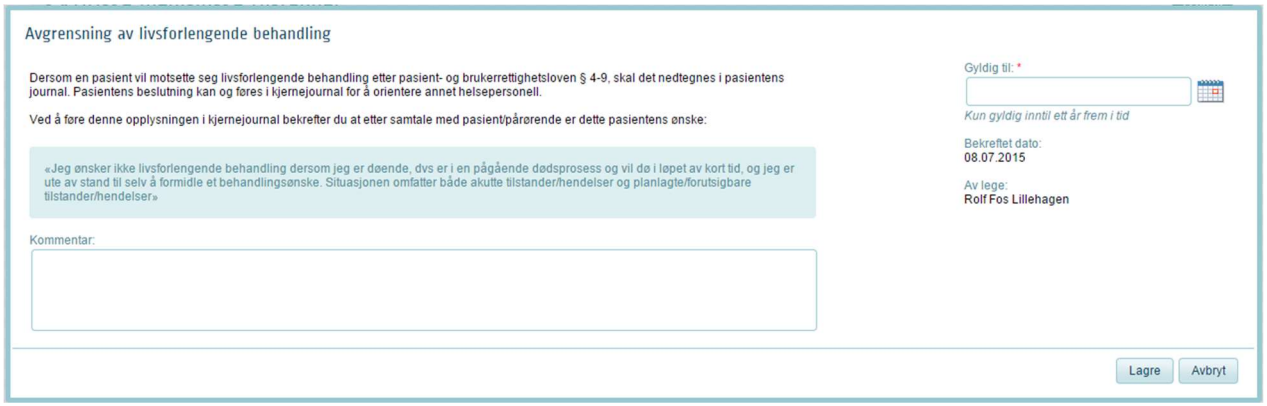


Figure 2-Recording in Emergency Care Summary Record

3.5.1.4 Validity period

Declaration that the desired treatment at the end-phase of life shall take place, with date. The health care personnel responsible for the treatment are those who shall assess whether the wish for treatment is still the patient's point of view. Entries in this category have a duration of 1 year, and may be renewed annually after discussion with the patient.

The reason for regularly renewing the declaration is to make sure the physician responsible for treatment is sure that the content still represents the patient's wish.

An entry in Summary Care Record is a warning to the treating doctor that the patient has previously asserted a position in respect of life-prolonging treatment in partnership with health care personnel.

The treating doctor must assess whether the patient's recorded wish for treatment applies to the current situation, or whether the patient may have changed their mind. The written statement shall constitute one of several sources of information when a decision needs to be made about reducing or withdrawing treatment.

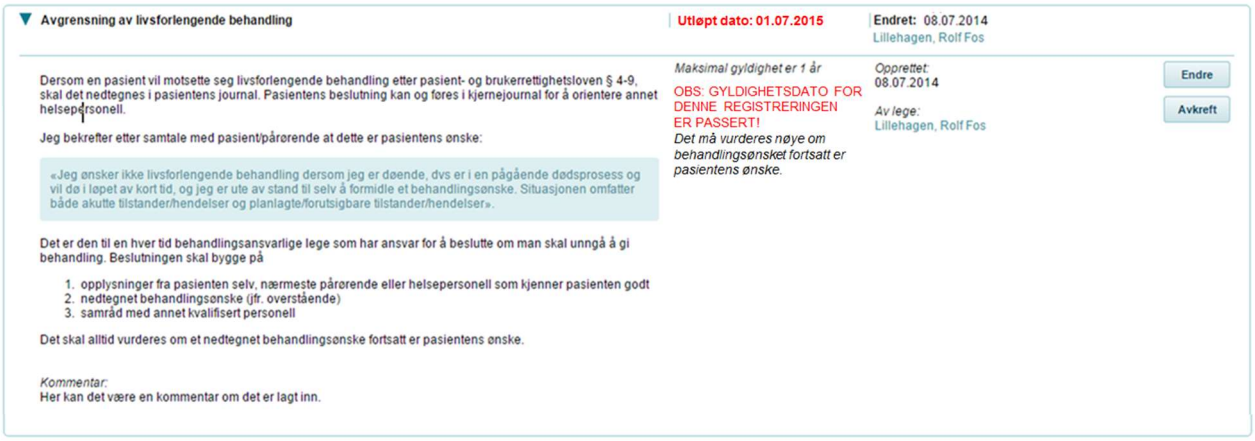


Figure 3- Detailed view of entry for which the validity date has expired.

3.5.2 *Patient who does not wish to undergo blood transfusion.*

Pursuant to the Act on Patient- and User Rights, § 4-9, paragraph one, a patient has an independent right to refuse to receive blood- or blood products based on a firmly-held belief. The choice must be made of their own free will, and must not be an expression of mental illness. It may be challenging to assess this, where a person within a religious milieu feels that this environment pressures or requires them not to receive blood.

Nonetheless, the patient has to right to have their view heard, and Summary Care Record can communicate this view. The need to record this information was identified early in piloting of Summary Care Record, and the reservation against blood transfusion has been entered in free-text fields intended for other purposes because no specific category for this was available.

It was also considered whether the patient could record this themselves in their own illness overview, but it was felt that this should be done in “Critical information” by the treating doctor, because then quality-assurance of the entry is made somewhat more secure. It provides some guarantee that the patient has not written this as an expression of mental illness, and the doctor can also, to some extent, make sure that the patient’s intended view is entered, and not something entered under pressure from other persons or milieus.
The information is entered under “Other procedural changes”.

3.5.3 *Other changes in procedure*

Other changes in treatment protocols, including limitations in treatment, are often linked to specific conditions, diagnoses or ongoing courses of treatment, which will be entered in one of the other categories in “Critical information”. Changes in a procedure can therefore be entered as a comment in the text box accompanying entry of the condition. The advantage of this approach is that everything that relates to a specific diagnosis or treatment is entered with it. The disadvantage of entering such information in the comments field is that one is not notified specifically that information about a change in treatment protocol has been entered. The care provider first sees this information on opening and reading what has been entered about the diagnosis or treatment. In general, nor is it desirable that the information is largely entered in an unstructured fashion in free text. It is therefore recommended that the changes in the treatment routine that should be notified be recorded in "Other procedural changes" as a separate sub-category under "Changes in normal treatment routine”.

3.5.3.1 *Other procedural changes - solution in Emergency Care Summary Record*

Description of the project change is done in free text, but grouped within one of the following categories:

1. The patient has stated they do not want to undergo blood transfusion.
2. Open admission agreement
3. Specific doctor/department should be contacted in the event of deterioration
4. Recommended specified treatment procedure
5. Other change in procedures

Validity period should be stated.

Figure 4 - Objection to blood products

3.5.3.1.1 *Specification of content in category "Other procedural changes":*

1. The patient has stated an objection to receiving blood transfusion/blood products
    a. Confirmation that the wish has been notified in person (mandatory)
    b. Expiry date (not mandatory)
    c. Expiry date (not mandatory)
2. Open admission agreement
    a. Specification of department/hospital concerned (mandatory)
    b. Comment: state condition and what has been agreed (mandatory)
    c. Expiry date (not mandatory)
3. Specific doctor/department should be contacted in the event of deterioration
    a. Specification of department/hospital concerned (mandatory)
    b. Comment: state condition and what has been agreed (mandatory)
    c. Expiry date (not mandatory)
4. Recommended specified treatment procedure
    a. Specification: which diagnosis/condition (mandatory)
    b. Comment: describe the treatment procedure (mandatory)
    c. Expiry date (not mandatory)
5. Other change in procedure
    a. Specification: which procedure (mandatory)
    b. Comment: describe procedural change (mandatory)
    c. Expiry date (not mandatory)

3.6 «Infection»

The “Infection” category contains notifiable infectious diseases only, where the disease has consequences for the choice of treatment for the patient. This applies in particular where the choice of antibiotics is crucial. There is no space for reporting diseases where the sole purpose is to inform health care personnel about possible risk of infection.

3.6.1 Limited information content

In treating the infected patient, it is important to know the nature of the infectious condition since it may affect the choice(s) of treatment. This group of patients often needs adapted treatment and despite the limited notification options in this category, it can be an important warning for other care providers. Among other reasons, use of antibiotics to which patients are resistant should be avoided since this can further enhance the infection.

Furthermore, there are cases where the patient has a known hepatitis (inflammation of the liver), where it is important to avoid drugs that may have potentially harmful side effects on the liver. Similarly, information about an active HIV-infection is important to prevent administration of pharmacotherapy that may further inhibit the patient’s immune system.

When an infected patient receives the treatment, it is important that precautions are taken to prevent spread of infection. But the purpose of the Summary Care Record is to warn about critical conditions significant for treatment of the specific patient, and infection control measures solely intended to protect others are not permitted as warnings in the Summary Care Record.

Information can also be given in Summary Care Record about infectious diseases where there is risk of the patient receiving the wrong treatment or being wrongly diagnosed, where the infection is unknown. This is stated by virtue of a general warning within the infection category (“Other infectious disease with consequences for treatment of the patient”), with the exceptions of MRSA, VRE and ESBL, which are notified separately. Other infectious diseases that will affect choice of patient treatment should be specified in more detail under “Medical condition”, “Ongoing treatment” or “Change in treatment procedure”, depending on what problems surround the condition. When the patient is infection free, the information should be removed from Summary Care Record.

3.6.2 Code values “Infection” in Summary Care Record’s critical info:

1. Disease caused by methicillin-resistant golden staphylococcus (MRSA)
2. Disease caused by vancomycin-resistant enterococcus (VRE)
3. Disease caused by extended-spectrum-beta-lactamases (ESBL)
4. Disease caused by other multi-resistant microbe
5. Other infectious disease with consequences for treatment of the patient

Figure5 - Registration of infection

3.7 Other non-structured information (converted from EPR)

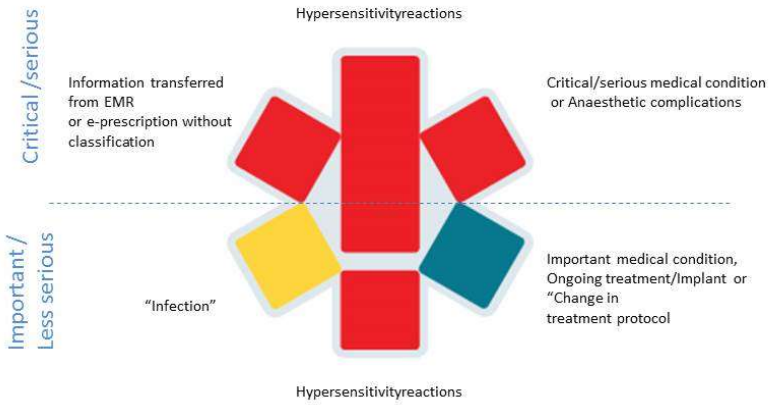
Current EPR-systems have no common, standardized structure for recording of critical information, and in many EPR-systems, entry of critical information occurs in free text. There is also varying data quality in the existing information. It is therefore unsafe to exchange this information with Summary Care Record without the information being verified and categorized by the treating doctor. The aim remains, nonetheless, to achieve integration between the current EPR and Summary Care Record so that critical information can be available across the systems.

A separate category has been defined in Summary Care Record for information awaiting approval by a doctor, but will not commence use until it is decided how close integration with EPR will occur. The category may, for example, be used when recording CAVE-information entered with ATC-codes from the e-prescription system.

4 Use of symbols in the Summary Care Record to alert critical information.

A symbol is displayed on all pages in Summary Care Record to signal that it contains critical information about the patient. Based on the model from the Swedish solution in NPÖ, a symbol has been chosen that can be described as a 6-armed cross, based on the “St. Andrew’s Cross” or “Star of Life”, without the Rod of Asclepius.

The information is notified as either **“Critical/Serious" (medical alert)** or **"Important / Less serious" (for observation/nota bene)** depending on the severity and category of the information**.**



It is therefore possible that the same type of information can be entered as "critical" or "important". CRITICAL/SERIOUS information will be signaled by marking the UPPER half of the symbol, while IMPORTANT/LESS SERIOUS information will be signaled by marking the LOWER half of the symbol.
Critical/Serious hypersensitivity reactions are presented in the central axis as an exclamation point, and just as a square at the bottom for important/less serious.

Critical/serious hypersensitivity reactions

Important/less serious hypersensitivity reactions

Entries under "medical condition" are indicated in red in the upper right arm, if the condition is on the “absolute list”, i.e. defined as serious, or lower right arm if it is another important diagnosis (colored in blue). Anesthetic complications are defined as “Critical/Serious” and are notified in red in the upper right arm

Critical/serious medical condition or Anesthetic complications

Important condition, Ongoing treatment/Implant or “Change in treatment protocol”

Entries in the categories “Change in treatment protocol” and “Ongoing treatment/Implant”, are signaled only as “important” in the right lower arm in blue

Information transferred from EMR or e-prescription without classification

"Other non-structural information” is indicated as critical in the upper left arm in red until this is assessed and categorized by the treating doctor.

“Infection”

“Infection is signaled in the lower left arm in yellow as “important”.

5 Code values in Summary Care Record

The following is a summary of the code values used in Summary Care Record:

5.1 Degrees of severity:

1. Severe reaction
   - Reaction that was potentially life-threatening or has caused injury to health with a given duration.
2. Less severe reaction
   - Reaction that was troublesome or unpleasant, but posed no risk to life or injury to health

5.2 Grading of probability (certainty):

1. *Suspected*
   All relationships that cannot be excluded, including when clinical observation is not uniform
2. *Probable*
   The connection is more likely than other possibilities, but there is no documentation describing it as “confirmed”
3. *Confirmed*
   The connection is documented by analyses, including provocation test, investigation by an allergologist or similar
4. *Disproven*
   Attempt made to confirm hypersensitivity reaction with laboratory tests or provocation tests, but these were negative and it is most probable that the patient does not suffer from these.

5.3 Source of information

1. *Information from the patient*
   Information from the patient, without the treating physician having been able to verify this objectively by tests or reliable documentation.
2. *Information from relatives*
   Information from relatives, where the patient cannot themselves verify this. Applies in particular to minors, and patients who are unable to self-report.
3. *Taken from existing notes*
   Information recorded in the EMR or other documents, without one knowing with certainty who the original source of information was, or where the person who

entered the information cannot be contacted where necessary to confirm the information.

4. *Stated by responsible care provider*
   The responsible care provider/health care personnel (doctor, psychologist, nurse etc.) can confirm that the information is correct. Often used when recording in categories other than hypersensitivity reactions, where one cannot say that the information is "observed" (critical diagnoses, treatments etc.). May also be used when the person entering the information has not personally observed a reaction, but is notified of it by another responsible health care worker with first-hand knowledge, e.g. via the discharge notes.
5. *Observed by treating doctor*
   Used primarily in the "hypersensitivity reactions" category, when the person entering the information about a reaction has also observed it.
6. *Result of tests/analyses*
   Used when a reaction is confirmed by analysis or tests (e.g. provocation testing). May also be used where there is an investigation and conclusion from an allergologist.
7. *Other*
   Other sources of information that do not naturally fit into one of the above code values. Comment with explanation should be given.

5.4 Shortlist of important allergies:

1. Milk
2. Eggs
3. Wheat
4. Shellfish
5. Fish
6. Peas
7. Soya
8. Lupine
9. Fenugreek seed
10. Peanuts
11. Nuts other than peanuts
12. Poppy seed
13. Insect toxin
14. Latex
15. Other important allergies (specified in free text)

Explanation/criteria:

The listed allergens are those commonly reported as causing *severe* allergic reactions, with the exception of pharmaceuticals. If using code 15 "Other important allergies", the allergen should be specified in free text. The allergen should be specified as precisely as possible. Avoid stating combination products containing numerous potential allergens. The allergy should be *severe* (see definition of severities) and important in the context of a medical acute.

5.5 Types of reaction:

- **Anaphylaxis**
  - Anaphylactic reaction (= Sudden onset reaction affecting at least 2 organ systems)
- **Reactions involving the circulatory system**
  - Hypotension
  - Serious arrhythmia
- **Reaction affecting the apparatus of breathing**
  - Laryngeal edema

- Asthma
- Unspecified difficulty breathing

- **Central nervous system reaction**
  - Seriously affected level of consciousness/confusion etc.
  - Generalized seizures
- **Reaction affecting skin/mucosa**
  - Angioedema/severe generalized urticaria
  - Other serious skin reactions such as Stevens-Johnson syndrome, epidermolysis, severe bullous reactions, vasculitis etc.
  - Less severe reactions such as itching/swelling/local urticaria/erythema
  - Irritation of the eyes, nose, throat
- **Reactions affecting the GI-tract**
  - Vomiting/diarrhea/abdominal pain
- **Other reactions**
  - Liver failure/hepatic impairment
  - Renal failure/renal impairment
  - Blood: aplasias/dysplasias/cytopenias
  - Rhabdomyolysis
  - Other severe reaction
  - Other less severe reaction
- **Unknown reaction**

Explanation/criteria:

Name	Degree of severity	Description
Anaphylaxis		
Anaphylactic reaction	S	Sudden onset reaction affecting 2 or more organ systems should be entered as anaphylaxis
Reaction involving the circulatory system		
Hypotension	S	Sudden drop in blood pressure that cannot be predicted on the basis of known pharmacological properties
Serious arrhythmia	S	Includes serious ventricular arrhythmias, cardiac arrest, serious AV-block and prolonged QT-interval
Reactions involving the respiratory system		
Laryngeal edema	S	Includes all acute swelling of the upper airway with obstruction
Asthma	S	Obstruction of the lower respiratory tract
Unspecified difficulty breathing	S	Other stridor/dyspnea, which cannot be classified as laryngeal edema or asthma (see above)
Central nervous system reaction		
Effects on level of consciousness/confusion	S	Includes unconsciousness. The reaction must have occurred on a dose of the medicinal product that does not normally cause such a reaction
Generalized seizures	S	Includes all serious extrapyramidal reactions. Seizures thought to be functional should not be entered
Reaction affecting skin/mucosa		
Angioedema/severe urticaria	S	Swelling of the face/lips/eyelids and/or generalized severe urticaria. Limited urticaria with no general effects are recorded as "less severe skin reaction"

Other severe skin reactions	S	Includes, among other things, Stevens-Johnson syndrome, epidermolysis, vasculitis, severe bullous skin diseases etc.
Less severe skin reaction	LS	Includes itching, local swelling, limited urticaria and erythema without constitutional symptoms
Irritation of the eyes, nose, throat	LS	Applies only to irritating reactions. Reactions causing obvious breathing difficulties are recorded “reaction affecting the respiratory system”
Reactions affecting the GI-tract		
Vomiting/diarrhea/abdominal pain	LS	Any gastrointestinal reactions, which cannot be anticipated based on the medicinal product’s known pharmacological properties
Other reactions		
Liver failure/hepatic impairment	S	Any gastrointestinal reactions, which cannot be expected based on the medicinal product’s known pharmacological properties
Renal failure/renal impairment	S	Effects on renal function, which cannot be anticipated based on the medicinal product’s known pharmacological properties at a normal dose.
Blood: Aplasias/dysplasias/cytopenias	S	All serious changes in the blood picture, which cannot be anticipated based on the medicinal product’s known pharmacological properties at a normal dose.
Rhabdomyolysis	S	Rapid breakdown of skeletal musculature, which cannot be anticipated based on the medicinal product’s known pharmacological properties at a normal dose
Other severe reaction	S	Other reaction that has been life-threatening or has caused serious damage to health. Remember to specify in free text
Other less severe reaction	LS	Another reaction that was distressing for the patient, but caused no serious harm to health. Remember to specify in free text

Degree of severity - indicative table (S = Severe LS = Less severe)

5.6 Intubation problem

- Grading of anatomical access for intubation (Cormack & Lehane)
    1. Grade 1: Vocal cords visible
    2. Grade 2: Posterior extremity of glottis seen
    3. Grade 3: Only epiglottis visible
    4. Grade 4: Epiglottis not visible
    5. Other intubation problems

- Able to mask ventilate?
    1. Yes, easily
    2. Yes, with difficulty
    3. No
    4. Not stated

5.7 Justification for critical condition

1. May affect level of consciousness
2. May have serious effects on respiration
3. May result in bleeding tendency
4. May cause circulatory failure
5. Risk of problems during anesthesia
6. Risk of complications during surgery
7. Risk of life-threatening complications if drug-treatment changes
8. Other (specify in free text)

Explanation/criteria:
Justification need only be stated when recording a diagnosis/condition that is NOT on “the absolute list”. It should state what the most serious problem is if the condition is overlooked in the context of a medical acute. Where the condition has potential to cause several problems, choose the most frequently occurring or most serious problem. Choose just one. Option 8 (other) should only be used in exceptional cases, where other, special problems apply.

5.8 Ongoing important treatment:

:

1. *Chemotherapy*
   Given at outpatient clinic or day ward, where the medication is not prescribed to the individual.
2. *Immunosuppressant/-modulating treatment*
   Given at outpatient clinic or day ward, where the medication is not prescribed to the individual.
3. *Radiotherapy*
   All forms of radiotherapy etc. given over time by regular visits to the outpatient ward, radiological day treatment, radiotherapy department or outpatient clinic.
4. *Important anticoagulant therapy*
   Drugs given to the patient on discharge to home treatment without a prescription. Can also be used to provide important additional information, not presented on the drug prescription/dispensing note (e.g.: “INR must be > 2.5. To have Klexane if INR < 2.5”)
5. *Dialysis*
   Al those needing regular dialysis
6. *DAR (Drug-assisted rehabilitation)*
   Patients for whom the DAR-treatment does not appear in the drug list (e.g. due to regular supply from outpatient department etc.)
7. *Participation in clinical pharmacological trials*
   Patients taking blinded drugs. Information about alternative active substances or placebo. Preferably contact number to break the code.
8. *Established psychiatric care plan*
   Patients with established crisis- or mastering plan. Entered in consultation with the patient.
9. *Ongoing outpatient care - pain clinic*
   Regular treatment at pain clinic where medication used does not appear in the drug list. For example, injections given without a prescription being written.
10. *Ongoing outpatient treatment - Chelate treatment*
    Outpatient chelate treatment given at hospital outpatient department or similar
11. *Ongoing investigation/follow-up of suspected or verified malignant disease*

Used when initiating outpatient (including general practitioners) investigations/treatment that does not come under the alternative codes above. Remember that this must be entered in consultation with the patient. Especially if the patient has pathological findings/results of analyses that are being followed-up. (To prevent doubled-up investigations).

12. Other outpatient treatment

Other important ongoing outpatient treatments that do not appear in the drug list.

13. On the transplantation waiting list

Patients who are accepted for transplantation and awaiting an organ.

14. Other important treatment

5.9 Implants

Head - neck - CNS

1. Ear implant (including cochlear implant)
2. Metal in the eye (foreign body, etc.)
3. Other implant in structures adjacent to the eye
4. Intracranial vascular clips, etc.
5. Shunt or similar in the central nervous system
6. Neurostimulator
7. Implant in the mouth, nose, throat
   Ordinary dental prostheses etc. should not be entered.
8. Tracheostomy
   Permanent tracheostomy should be entered

Cardiovascular

9. Pacemaker
   Permanent pacemaker should be entered. Enter type and if possible, MRI-compatability
10. Implanted defibrillator (ICD)
11. Mechanical heart valve
12. Other implanted heart valve
13. Coronary stent
    Should be entered when the patient is receiving double anticoagulation therapy (9–12 months after implantation). Older stents are of interest only in the context of medical history and need not be entered.
14. Stent in other than coronary vessels
15. Vascular prosthesis

Abdomen

16. Stent, catheter or implant in the upper urinary tract
    Ordinary urinary catheter is NOT entered

17. Biliary stent
18. Stent in other hollow organ
19. PEG-tube

Pumps

20. Infusion pump/syringe driver
    (Electronic medical pump or other injection/infusion equipment)

Orthopedic

21. Orthopedic prosthesis or implant
    (Only prosthesis or implant significant in the context of an acute medical situation)

Transplants

22. Transplanted kidney
23. Transplanted kidney
24. Transplanted lung
25. Transplanted heart and lungs
26. Transplanted liver
27. Transplanted bone marrow, pancreas, intestine, stem cells
28. Other transplanted organ or tissue

Other

29. Other implant
    (Other implant, foreign object, prosthetic or aid worn on the body)

***General comments*:** Only implants of significance in the context of a medical acute should be recorded. Most are self-explanatory and not commented on specifically.

5.10 Procedural changes

1. Limitation of life-prolonging treatment
2. Other changes in procedure
    a. The patient has stated they do not want to undergo blood transfusion.
    b. Open admission agreement
    c. Specific doctor/department should be contacted in the event of deterioration
    d. Recommended specified treatment procedure
    e. Other change in procedures

Explanation/criteria:

Limitation of life-prolonging treatment is regulated by laws and regulations. See description in the report.

5.11 Code values for “Infection” in Summary Care Record’s critical info:

1. Disease caused by methicillin-resistant golden staphylococcus (MRSA)
2. Disease caused by vancomycin-resistant enterococcus (VRE)
3. Disease caused by extended-spectrum-beta-lactamases (ESBL)
4. Disease caused by other multi-resistant microbe
5. Other infectious disease with consequences for treatment of the patient

Explanation / criteria:

Only the specified multi-resistant microbes are specifically indicated. Only contagious disease where the knowledge of the infection will lead to a change in the patient's medical treatment shall be stated. Infection information that will only entail protective measures for health personnel should not be disclosed. When using Code 5 " Other infectious disease with consequences for treatment of the patient ", details should be provided by an entry under "Critical Medical Conditions", "Ongoing Treatments" or " Changes in treatment protocol" depending on what is the consequence of the condition.

6 Sample images from Critical Info in Summary Care Record

Below are some sample images from the pilot version of National Emergency Care Summary Record:

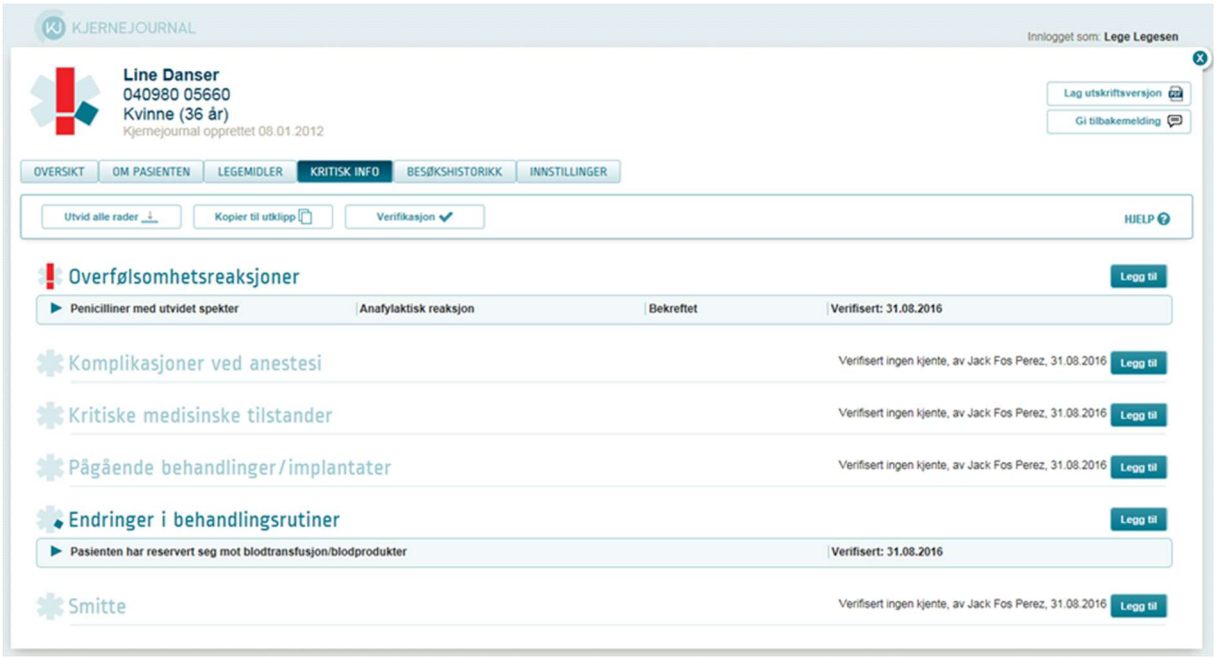


Figure 7. Critical info - Main image with 2 active categories

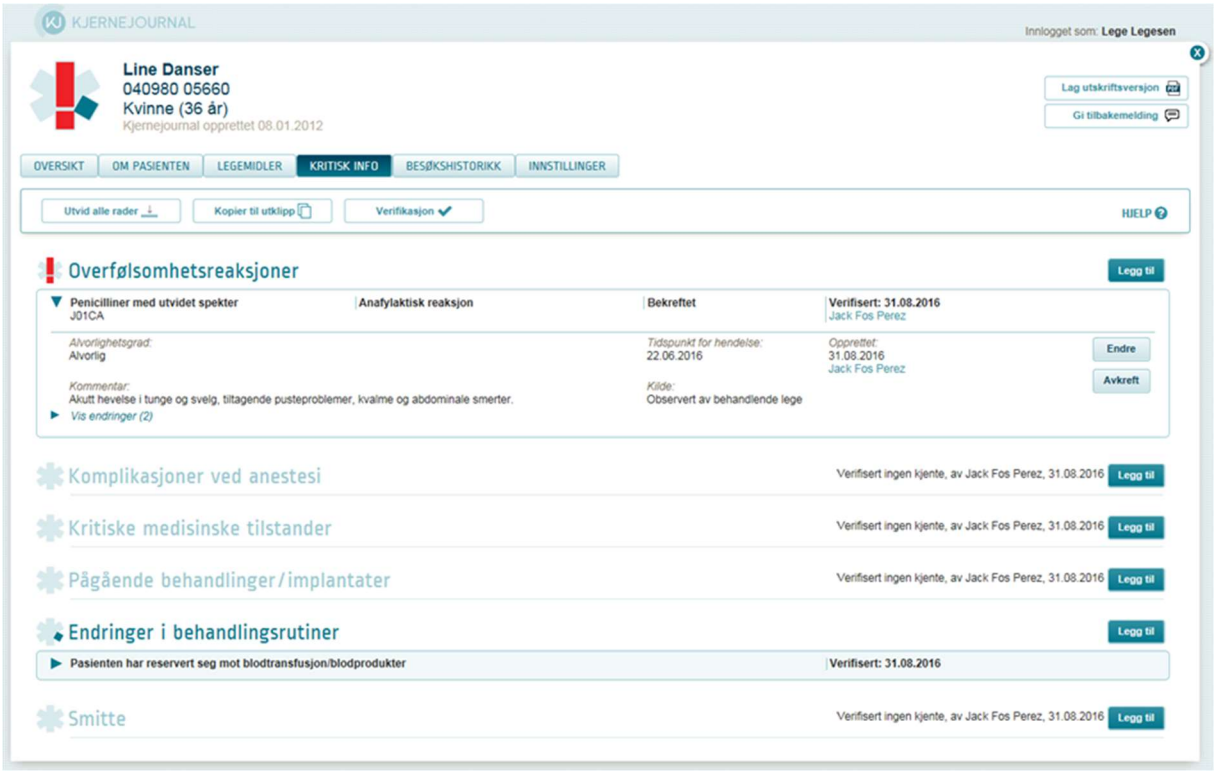


Figure 8. Detailed View of entered hypersensitivity reaction

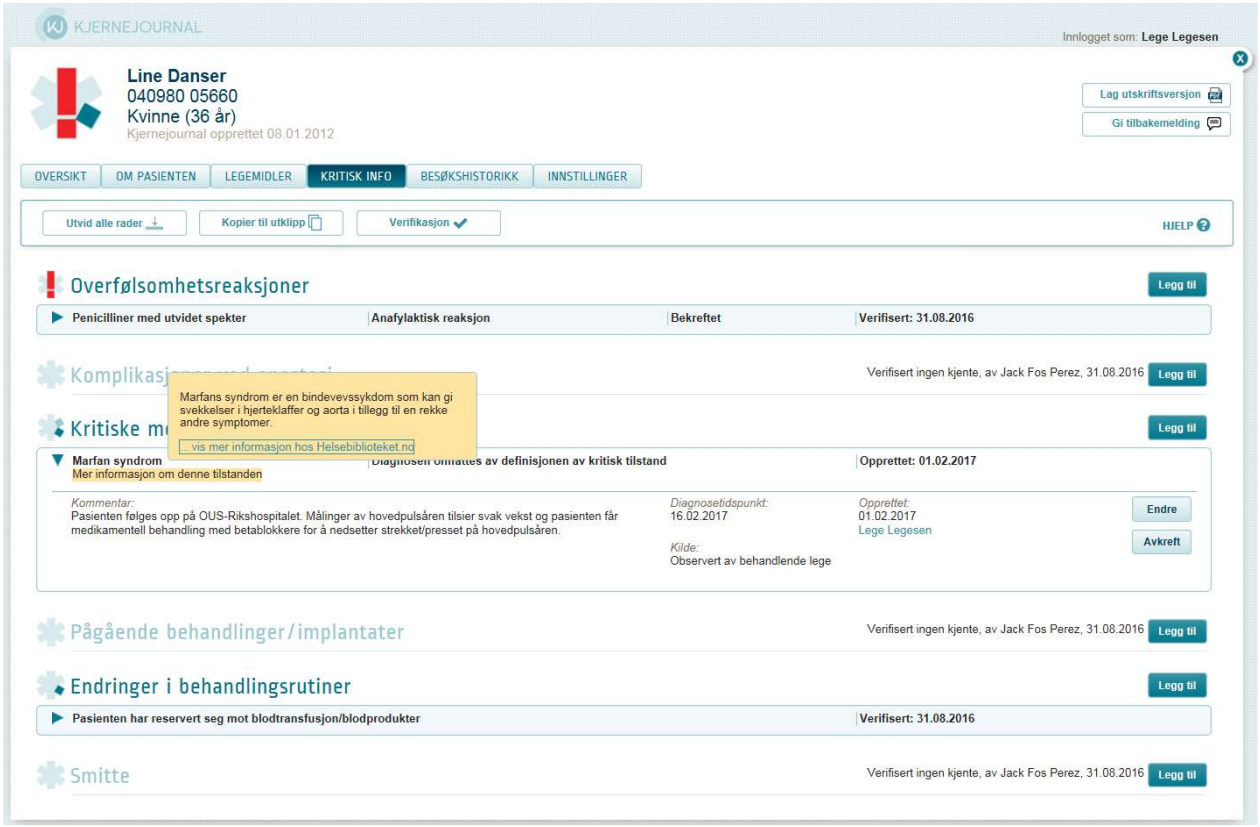


Figure 9. Detailed view of Critical medical condition with information

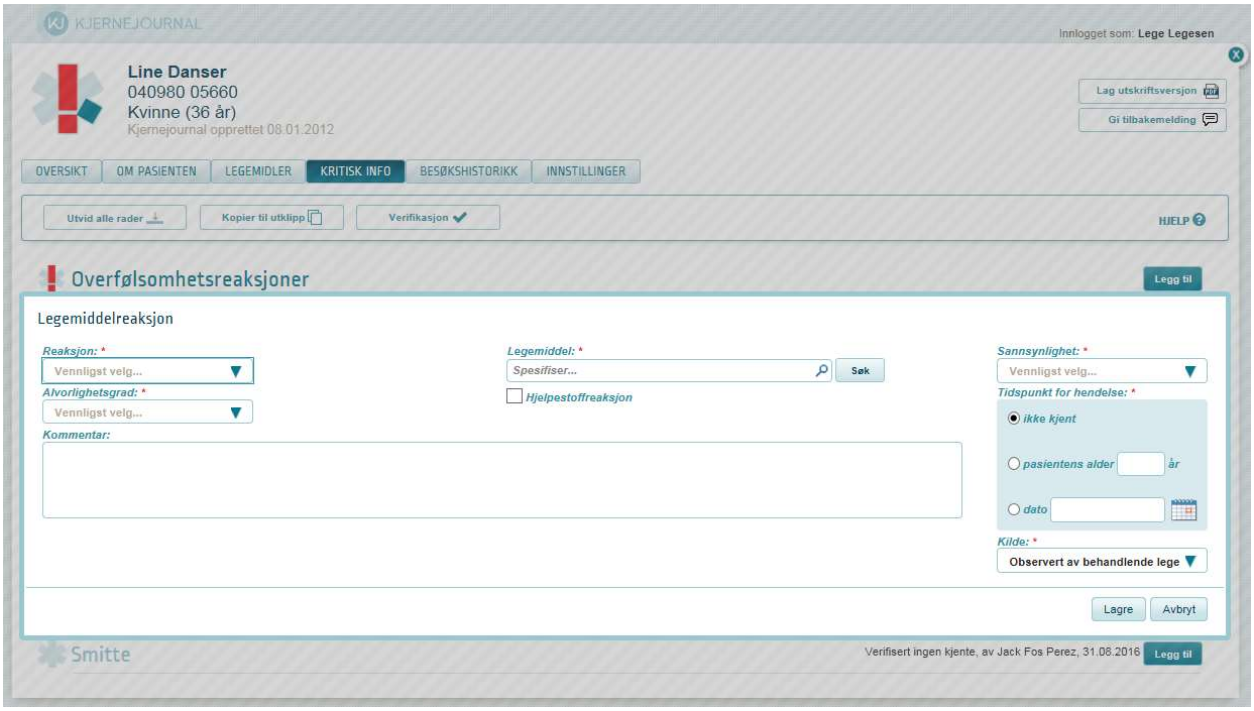


Figure 10 Registration image for new drug reaction

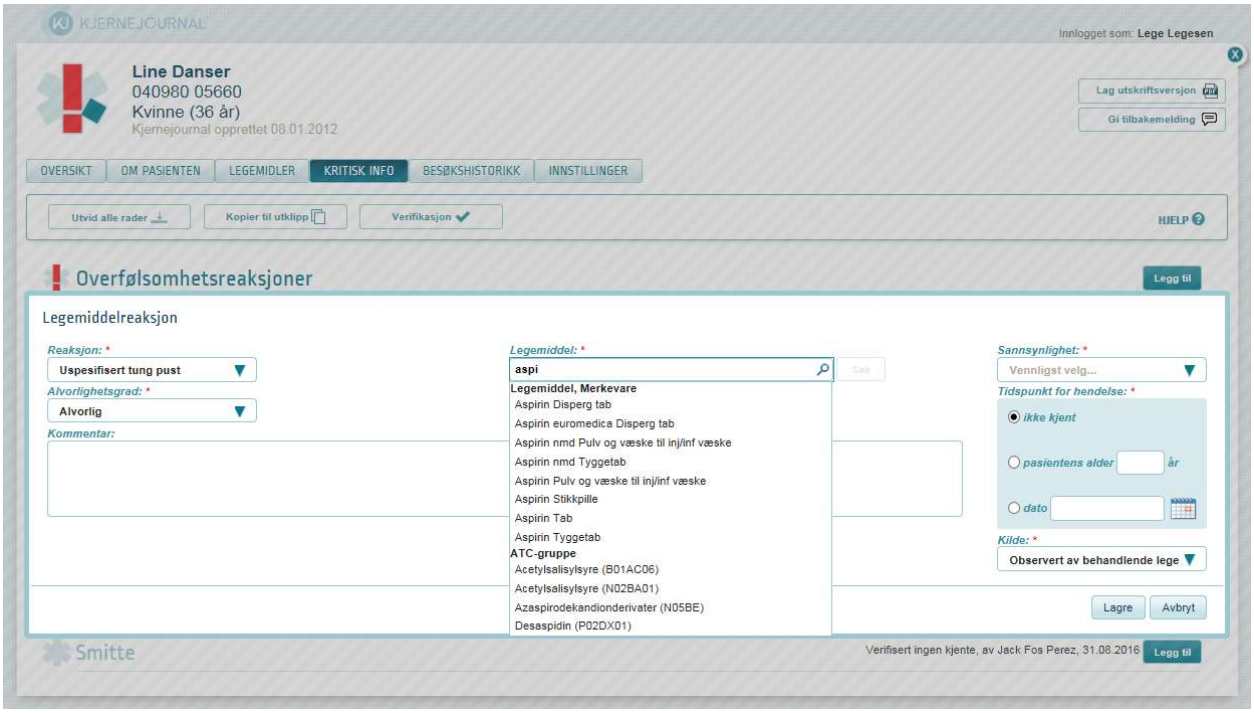


Figure 11 Registration of drug reaction - Search based on drug/ATC-code

Enclosure A

List of diagnoses/conditions that should be registered as “Critical medical condition” in Summary Care Record (“the absolute list”), as of 1.july 2018.

6.1 Appendix 1 - The absolute list

List of diagnoses / conditions that should be recorded as "Critical medical condition" in SCR. This is a dynamic list that is continually updated and adapted to actual clinical needs in line with current knowledge and practice. The list is updated as of June 2018

The list consists of:

- "Term" is the normal Norwegian or international name (when Norwegian name does not exist or is not adequate)..
- "Keywords" are other terms that are used about the condition, and also "popular" terms. Also included are ICPC codes that include the condition.
- "Matching ICD-10 codes" are the ICD-10 code (s) that classify the state
- "Rationale" is a brief description of why the condition is included as "critical". The description also gives some brief advice on how the patient should be handled in an emergency situation.

There are 2 "levels" of diagnoses / conditions in the absolute list. We have gathered together a number of conditions with approximately equal consequences for treatment choices, in groups to simplify the registration in the SCR. Examples of such groups are "blood clot tendency", "muscular dystrophy", "platelet defect" etc. These are conditions with approximately equal consequences for the treatment and therefore it is often not necessary to specify precisely which specific diagnosis is concerned. Within each group, however, one or more more specified conditions are indicated which can be specified instead of the group term if desired.

6.1.1 Overview of collective terms

Hemophilia

- Hemophilia A (Classic hemophilia)
- Hemophilia B

Epidermolysis bullosa (EC)

- Herlitz syndrome

Disturbance of fatty acid metabolism

- MCAD
- LCHAD
- CPT 1
- CPT 2

Hypopituitarism

- pituitary tumors
- Kallmann's syndrome
- Lorain-Levi dwarfism
- Sheehan's syndrome
- Simmond's disease
- Craniopharyngioma

Immunodeficiency

- AIDS

Long QT syndrome

- Jervell-Lange-Nielsen
- Andersen – Tawil syndrome
- Romano – Ward syndrome

Mucopolysaccharidosis

- Hurler's disease (MPS I)
- Hunter's disease (MPS II)
- Sanfilippo's disease (MPS III)
- Morquio's disease (MPS IV)
- Maroteaux-Lamey's disease (MPS VI)
- Accidental disease (MPS VII)
- MPS IX

Muscular / myopathies

- Pumpe's disease
- Emery-Dreifuss muscular dystrophy
- Duchenne muscular dystrophy
- Becker muscular dystrophy
- Myotonic dystrophy (Steinert's disease)
- Thomsen's disease

Organic aciduria

- Glutaric Acidaemia

Pulmonary hypertension

- Eisenmenger syndrome

Platelet Defect

- Thrombocytopenic purpura
- Thrombocytopenia
- Scott's syndrome
- Storage pool deficiencies

Trombocytopati

- Glanzmann's thrombasthenia
- Bernard-Soulier's syndrome
- Idiopathic thrombocytopenia (ITP)

Blood clots Tendency

- thrombophilia
- Antikardiolipinsyndrom
- Antiphospholipid
- Presence of lupus anticoagulant
- Activated protein C resistance
- Factor V Leiden mutation
- antithrombin deficiency
- Protein C deficiency
- Protein S deficiency
- Protrombingenmutasjon

Urea Cycle Defects

- Argininemi
- Arginino-suksininsyreuri
- Ornithine transcarbamylase deficiency (OTC)
- Citrullinemi

Hyperammonaemia

Vascular malformations in the brain

- Moyamoya

Term	Norwegian synonyms/key words	Matching ICD-10 codes	Rationale
AIDS	HIV-sykdom/AIDS	B24 B21 B20 B22	Patients often have serious infections masked as ordinary infections. Require special vigilance and prevention of infection following surgical procedures.
Activated protein C-resistance	Trombofili		Patients with an abnormal tendency to form blood clots. Blood clots may stick in the lungs and brain and cause life-threatening complications. Diagnostic vigilance is required, and special precautions must be implemented in a variety of treatments.
Adrenocortical insufficiency	Binyrebarksvikt Medfødt binyrebarksvikt CAH Addisons sykdom Cortisolmangel Kortisolmangel Mb. Addison Morbus Addison	E27.1 Primary adrenocortical insufficiency	Adrenocortical insufficiency results in a lack of the stress-hormone cortisol, and is usually treated with substitution therapy with cortisol tablets. Where levels are low, a variety of symptoms appear such as psychosis, convulsions and coma and at worst life-threatening shock accompanied by hypoglycemia, electrolyte disturbances and more. Dose adjustments are required for physical and psychological stress, e.g. surgery.
Alport syndrome	Hereditær nefritt Arvelig nefritt Q87.8		Alport syndrome is a rare and hereditary syndrome characterized by kidney disease and hearing loss that develops over time. Sometimes vision is also affected. In some cases, kidney disease causes severe renal failure, which must be treated with dialysis and / or kidney transplantation.

Amyloidosis	Familiær middelhavsfeber Amyloid polynevropati (Portuguese)	E85 Amyloidosis E85.0 Familial hereditary non-neurogenic amyloidosis E85.1 Familial hereditary neurogenic amyloidosis E85.2 unspecified familial hereditary amyloidosis E85.3 Secondary systemic amyloidosis E85.4 Organ-limited amyloidosis E85.8 Other specified amyloidosis E85.9 unspecified amyloidosis	Amyloidosis is a condition involving deposits of protein in various organs as a result of inflammatory conditions. Symptoms vary depending on the organ affected, but the disease may cause bleeding and organ failure with symptoms stemming respectively from the kidneys, liver and heart.
Andersen-Tawil syndrome	Hjertearytmi		A congenital conduction disturbance in the heart causing the patient to be at risk of life-threatening arrhythmia. Patients must avoid a number of medicinal products that prolong the QT-interval.
Angioedema	Non histamine-induced angioedema Angionevrotisk ødem Angioneurotisk ødem Non-histamin angioødem Quinckes ødem	T78.3 Angioneurotic edema	The patient may develop acute swelling of the dermis and mucosa of the face and upper respiratory tract. May have a turbulent course, and respiratory failure may result if treatment is not commenced.
Antifosfolipidsyndrome	Trombofili		Patients with an abnormal tendency to form blood clots. Blood clots may stick in the lungs and brain and cause life-threatening complications. Diagnostic vigilance is required, and special precautions must be implemented in a variety of treatments.
Antikardiolipin-syndrome	Trombofili		Patients with an abnormal tendency to form blood clots. Blood clots may stick in the lungs and brain and cause life-threatening complications. Diagnostic vigilance is required, and special precautions must be implemented in a variety of treatments.

Antitrombindeficiency	Trombofili		Patients with an abnormal tendency to form blood clots. Blood clots may stick in the lungs and brain and cause life-threatening complications. Diagnostic vigilance is required, and special precautions must be implemented in a variety of treatments.
Aortic aneurysm	Aneurisme under oppfølging Abdominalt aortaaneurisme Torakalt aortaaneurysme Thorakalt aortaaneurisme	I71.2 Thoracic aortic aneurysm without mention of rupture I71.4 Abdominal aortic aneurysm without mention of rupture I71.6 Thoracoabdominal aortic aneurysm without mention of rupture	Aortic aneurysms monitored conservatively, or awaiting treatment, can also rupture. This can be difficult to recognize, and information about the condition may increase the chance of a rapid diagnosis.
Argininemia			Metabolic disease where the patient often follows a low-protein diet with amino acid supplements. The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral nutrition.
Arginino-suksininsyreuria			Metabolic disease where the patient often follows a low-protein diet with amino acid supplements. The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral nutrition.

Bardet-Biedl syndrome (BBS)	Laurence-Moon-Bardet-Biedl syndrom (LMBB)		LMBB/BBS is a rare congenital syndrome with a wide spectrum of clinical findings such as obesity, retinitis pigmentosa (degeneration of the retina), polydactyly (redundant fingers/toes), delayed development, and decreased kidney function. Other problems may include oral/dental problems, heart problems, asthma, learning disabilities, speech problems, mental disorders and diabetes mellitus. In acute situations, be particularly observant for possible renal failure and heart problems
Bartter syndrome	hypokalemi metabolsk alkalose hypomagnesemi Primary renal tubular hypokalemic alkalosis Hyperprostaglandin E syndrome		Bartter syndrome is a rare genetic disorder that causes low levels of potassium in the blood. Symptoms may include paraesthesia, paralysis or convulsions. Patients are dependent on potassium replacement therapy, and conditions such as vomiting, diarrhea and fever can worsen symptoms and lead to an increased need for substitution.
Bernard-Souliers syndrome	Blodplatedefekt Purpura		Lack of- or dysfunctional platelets results in delayed coagulation of the blood, such that the patient may die from blood loss or organ damage. Particular caution should be observed during surgery and with acute injuries.
Confirmed blood group antibodies	Erytrocyttantistoff Blodtypeantistoff Trombocyttantistoff	R76.0 Elevated antibody titers T80.3 ABO incompatibility reaction T80.4 Rh incompatibility reaction T80.5 Anaphylactic shock due to serum treatment T80.6 Other serum reactions T80.8 Other complications following infusion, transfusion and	Patients with known blood group antibody must receive special intervention on blood transfusion. Even transfusion with “crisis blood” (o Rh-) can cause life-threatening haemolysis in patients who have circulating erythrocyte antibodies.

		therapeutic injection	
CADASIL	Cerebral Autosomal-Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalo-pathy CADASIL syndrome		A hereditary disorder affecting the blood vessels in the brain. Involves very high probability of stroke at a young age.
Carnitine deficiency	Carnitinmangel	E71.3 Disorders in the metabolism of fatty acids	Carnitine deficiency is a deadly, congenital disorder in the conversion of fatty acids, resulting, among other things, in cardiomyopathy, hypoglycaemia and liver failure. Patients must have carnitine-substitution treatment for the rest of their life. Increased risk in anesthesia.
CHARGE syndrome	Q87.8		Congenital genetic disorder. Emergency medical consequence: observe possible midline defects in intubation, skeletal anomalies; Note on hyperextension of the neck
Citrullinemia			Metabolic disease where the patient often follows a low-protein diet with amino acid supplements. The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral nutrition.
Cleidocranial dysplasia (CCD)			Congenital hereditary defect in skeletal development, characterized by incomplete or absent collarbone, delayed closure of the fontanelles and skull sutures, together with various dental problems. The condition causes aberrant

			head- and facial features and often short stature. Mental development is normal. NOTE: acute situation: Increased risk of atlanto-occipital/axial dislocation on intubation!
CPT 1			Metabolic disease The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral glucose polymer or parenteral nutrition containing 12-15% glucose via the nasogastric route. Lipids must not be administered intravenously.
CPT 2			Metabolic disease The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral glucose polymer or parenteral nutrition containing 12-15% glucose via the nasogastric route. Lipids must not be administered intravenously.
Craniopharyngioma			Hypopituitarism leads to failure in the production of one or more hormones. The symptoms can vary greatly, including degrees of adrenal failure, hypothyroidism, diabetes insipidus and more. One must be aware of electrolyte disturbances and the need for hormone substitution.

Cystic fibrosis	Cystisk fibrose CF	E84 Cystic fibrosis	Hereditary disease causing chronic cough with tenacious, purulent expectorate and symptoms of chronic sinusitis, together with altered bowel pattern, malnutrition and reduced general health. Retarded growth and development (late puberty). NOTE: acute situation: May have serious effects on respiration.
Deaf-blindness	Blindhet Døvhet/hørselstap		Causes significant communication challenges for the patient, and can be difficult to distinguish from other disorders, e.g. stroke.
Di Georges syndrom		D82.1 Di Georges syndrom	Congenital disorder. Emergency medical consequence: observe possible midline defects in intubation, skeletal anomalies (be aware in hyperextension of the neck), hypocalcaemia, immune deficiency.
Disorder of fatty-acid metabolism	CPT 1 CPT 2 Fettsyreoksydasjons defekt Forstyrrelse i fettsyrestoffskifte LCHAD MCAD	F71.3 Disorder of fatty-acid metabolism	Metabolic disease The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral glucose polymer or parenteral nutrition containing 12-15% glucose via the nasogastric route. Lipids must not be administered intravenously.
Dissociative and conversion disorder	Dissosiative lammelser Funksjonelle lammelser Funksjonelle kramper PNES	F44.9 Dissociative and conversion disorder	Dissociative disorders are found in several variants, and primarily involve a lack of linkage between symptoms and cause, will and function (eg lack of gait function even if physics works properly). The most common is "PNES" (psychogenic non-epileptic seizures) and conditions with

			paralysis and emotional disturbance of non-organic cause. Psychogenic non-epileptic seizures are epilepsy-like seizures that are believed to have psychological causes. Many patients with such seizures receive an erroneous epilepsy diagnosis and many years of antiepileptic medication. The attacks often represent major diagnostic and therapeutic challenges.
Dravet's syndrome	Epilepsi Dravets syndrom Generalized epilepsy with febrile seizures plus GEFS+		Dravet's syndrome is a serious, complex form of epilepsy occurring in children and adolescents.
Dysphagia	PEG-sonde Svelgvansker Svelgparalyse Dysfagi Svelgparese	R13 Dysphagia	Dysphagia or severe swallowing difficulties may result in food or liquid going down into the patient's airways. This may, again, result in life-threatening situations. Some such patients have a tube inserted surgically directly into the stomach ("PEG-tube").
Dystrofia myotonica	Steinerts sykdom		Muscular diseases can affect respiration after anesthesia. May also have undiagnosed cardiomyopathy or conduction disturbances. Requires individual investigation prior to anesthesia. Increased effects of muscle relaxants than others, and also increased risk of developing malignant hyperthermia requires special choice of anesthetic agents.
Ehlers-Danlos syndrome		Q79.6 Ehlers-Danlos syndrome	Congenital connective tissue disease with significant risk of dilatation of- or acute bleeding from major arteries, ruptured bowel or ruptured uterus during pregnancy.

Eisenmenger syndrome			Eisenmenger syndrome is pulmonary hypertension caused by congenital malformations of the heart. The patients have heart failure, respiratory failure and may experience a tendency to both bleeding and clotting.
Epidermolysis bullosa (EB)		Q81.0 Epidermolysis bullosa simplex Q81.2 Q81.8 Q81.9	Hereditary skin disease characterized by blister formation in various layers of the skin. Some severe forms may also involve risk of mucosal injury on intubation, catheterization etc. Do not use tape/plasters on the skin!
Fabry disease	Anderson-Fabrys sykdom Morbus Fabry angiokeratoma corporis diffusum Mb. Fabry Sphingolipidosis E75.2		An enzyme defect that can have serious effects on CNS and the heart. Patients may suffer episodes of severe pain.
Faktor V Leidenmutation	Trombofili		Patients with an abnormal tendency to form blood clots. Blood clots may stick in the lungs and brain and cause life-threatening complications. Diagnostic vigilance is required, and special precautions must be implemented in a variety of treatments.
Fatty acid oxidation defect	MCAD LCHAD CPT 1 CPT 2	E71.3 Disorders in the metabolism of fatty acids	Metabolic disease The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral glucose polymer or parenteral nutrition containing 12-15% glucose via the nasogastric route. Lipids must

			not be administered intravenously.
Fibrodysplasia ossificans progressiva	Myositis ossificans Stone Man Syndrome FOP Progrediererende myositis ossificans Progressiv fibrodysplasi	M61.1 Myositis ossificans progressiva/Fibrodysplasia ossificans progressiva	A connective tissue disease where damaged muscle tissue is converted to bone instead of healing, e.g. following intramuscular injections and surgery. May involve intubation problems and cardiovascular problems.
Galactosaemia	Galaktokinase-mangel	E74.2 Disorders in galactose metabolism	Enzyme deficiency requiring a milk-free diet. Often have expressive language/speech problems, which can cause communication problems. CAUTION: when using enteric feed. Drugs containing lactose/lactose should be avoided where possible
Genetic deviation in CYP metabolism – unspecified	farmakogenetikk CYP2C9 CYP2C19 CYP2D6 Cytokrom p450		Detected genetic abnormalities in the CYP enzyme system may affect the metabolism of a variety of drugs. There are gene variants that can both increase and decrease serum levels of several drugs. If a genetic deviation has been demonstrated, this should be considered before starting treatment with drugs that are degraded or activated via the CYP system.

Genetic deviation in CYP metabolism – CYP2C19 *17/*17			Ultra Rapid metabolisers: The genotype indicates increased metabolism and increased dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated via CYP2C19 (eg, clopidogrel), this genotype will increase the risk of side effects.
Genetic deviation in CYP metabolism – CYP2C19 *2/*2			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2C19 (eg, clopidogrel), this genotype will produce reduced efficacy.
Genetic deviation in CYP metabolism – CYP2C19 *2/*3			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2C19 (eg, clopidogrel), this genotype will produce reduced efficacy.
Genetic deviation in CYP metabolism – CYP2C19 *2/*4			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2C19 (eg, clopidogrel), this genotype will produce reduced efficacy.
Genetic deviation in CYP metabolism – CYP2C19 *3/*3			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2C19 (eg, clopidogrel), this genotype will produce reduced efficacy.

Genetic deviation in CYP metabolism – CYP2C19 *3/*4			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2C19 (eg, clopidogrel), this genotype will produce reduced efficacy.
Genetic deviation in CYP metabolism – CYP2C19 *4/*4			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2C19 (eg, clopidogrel), this genotype will produce reduced efficacy.
Genetic deviation in CYP metabolism – CYP2C9 *3/*3			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2C9 this genotype will produce reduced efficacy.
Genetic deviation in CYP metabolism – CYP2D6 *1/*1 >2 gencopies			Ultra Rapid metabolisers: The genotype indicates increased metabolism and increased dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated via CYP2C19 (eg, clopidogrel), this genotype will increase the risk of side effects.
Genetic deviation in CYP metabolism – CYP2D6 *3/*3			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2D6 this genotype will produce reduced efficacy.
Genetic deviation in CYP metabolism – CYP2D6 *3/*4			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2D6 this genotype will produce reduced efficacy.

Genetic deviation in CYP metabolism – CYP2D6 *3/*41			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2D6 this genotype will produce reduced efficacy.
Genetic deviation in CYP metabolism – CYP2D6 *3/*5			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2D6 this genotype will produce reduced efficacy.
Genetic deviation in CYP metabolism – CYP2D6 *3/*6			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2D6 this genotype will produce reduced efficacy.
Genetic deviation in CYP metabolism – CYP2D6 *4/*4			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2D6 this genotype will produce reduced efficacy.
Genetic deviation in CYP metabolism – CYP2D6 *4/*41			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2D6 this genotype will produce reduced efficacy.
Genetic deviation in CYP metabolism – CYP2D6 *4/*5			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2D6 this genotype will produce reduced efficacy.

Genetic deviation in CYP metabolism – CYP2D6 *4/*6			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2D6 this genotype will produce reduced efficacy.
Genetic deviation in CYP metabolism – CYP2D6 *5/*41			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2D6 this genotype will produce reduced efficacy.
Genetic deviation in CYP metabolism – CYP2D6 *5/*5			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2D6 this genotype will produce reduced efficacy.
Genetic deviation in CYP metabolism – CYP2D6 *5/*6			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2D6 this genotype will produce reduced efficacy.
Genetic deviation in CYP metabolism – CYP2D6 *6/*41			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2D6 this genotype will produce reduced efficacy.
Genetic deviation in CYP metabolism – CYP2D6 *6/*6			Poor metabolizers: The genotype indicates no metabolism and lower dosage requirements of drugs that are degraded by this enzyme. For prodrugs activated by CYP2D6 this genotype will produce reduced efficacy.

Gitelman's syndrome	hypokalemi metabolsk alkalose hypomagnesemi Primary renal tubular hypokalemic alkalosis Hyperprostaglandin E syndrome		Gitelman's syndrome is a rare genetic disorder that causes low levels of potassium and magnesium in the blood. Symptoms may include paraesthesia, paralysis or convulsions. Patients are dependent on potassium-replacement therapy, and conditions such as vomiting, diarrhea and fever can worsen symptoms may lead to an increased need for substitution.
Glanzmanns trombastenia	Blodplatedefekt		Dysfunctional platelets results in delayed coagulation of the blood, such that the patient may die from blood loss or organ damage. Particular caution should be observed during surgery and with acute injuries.
Glutarsyreuria			Metabolic disease where the patient often follows a low-protein diet with amino acid supplements. The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral nutrition.
Glycogen storage disease	Glykogen GSD Pompes sykdom Glykogenlagrings-sykdom Glykogenoser	E74.0 Glycogen Storage Disease	Metabolic disease where the patient often follows a low-protein diet with amino acid supplements. The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral glucose polymer or parenteral nutrition containing

			10% glucose via the nasogastric route.
Hemophilia	Hemofili Koagulasjonsdefekt Von Willebrands sykdom	D66 Hereditary factor VIII deficiency D67 Hereditary coagulation factor deficiency IX D68.0 Von Willebrand disease D68.1 Hereditary coagulation factor deficiency XI D68.2 Hereditary lack of other clotting factors	Hemophilia results in delayed coagulation of the blood, meaning that patients may die from blood loss after organ injury. Particular caution should be observed during surgery and with acute injuries. Emergency surgery should be performed in consultation with the hematological diseases unit at Rikshospitalet
Herlitz' syndrome		Q81.1 Epidermolysis bullosa letalis	Hereditary skin disease characterized by blister formation in various layers of the skin. Some severe forms may also involve risk of mucosal injury on intubation, catheterization etc. Do not use tape/plasters on the skin!
Hunter Syndrome (MPS II)		E76.1 Mukopoly-saccharidosis, type II	Mucopolysaccharidoses are hereditary, congenital metabolic diseases caused by defective degradation of mucopolysaccharides. Acute situation/anesthesia: Atlantoaxial instability occurs. Risk of crushing the medulla.
Huntington chorea		G10 Huntingtons disease	
Hurlers syndrome (MPS I)		E76.0 Mukopoly-saccharidosis, type I	Mucopolysaccharidoses are hereditary, congenital metabolic diseases caused by defective degradation of mucopolysaccharides. Acute situation/anesthesia: Atlantoaxial instability occurs. Risk of crushing the medulla.

Hydrocephalus	Vannhode	G91 Hydrocephalus G91.0 Communicating hydrocephalus G91.1 Obstructive hydrocephalus G91.2 Low pressure hydrocephalus G91.3 unspecified posttraumatic hydrocephalus G91.8 Other hydrocephalus G91.9 Hydrocephalus, unspecified	Hydrocephalus involves high intracranial pressure. A number of common symptoms may be a sign of life-threatening herniation.
Hyperammonemia			Metabolic disease where the patient often follows a low-protein diet with amino acid supplements. The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral nutrition.
Hypopituitarism	hypofysesvulst Kallmanns syndrom Lorain-Levi-dvergvekst Panhypopituitarisme Sheehans syndrom Simmonds sykdom	E23 Hypofunction and other disorders of the pituitary gland E23.0 hypopituitarism E23.2 Diabetes insipidus	Hypopituitarism leads to failure in the production of one or more hormones. The symptoms can vary greatly, including degrees of adrenal failure, hypothyroidism, diabetes insipidus and more. One must be aware of electrolyte disturbances and the need for hormone substitution.
Hypoparathyroidism	Hypoparathyreoidisme Hypoparathyreoidismus	E20 Hypoparathyroidism E20.0 Idiopathic hypoparathyroidism E20.1 Pseudohypoparatyreoidism E20.8 Other hypoparathyroidism E20.9 Hypoparathyroidism, unspecified	Lack of parathyroid hormone, a hormone that maintains the correct level of calcium in the blood. Calcium deficiency may, among other things, cause serious cardiac rhythm disturbances and respiratory failure in addition to a series of other symptoms.

Immune thrombocytopenic purpura		D69.3 Immune thrombocytopenic purpura	Lack of- or dysfunctional platelets results in delayed coagulation of the blood, such that the patient may die from blood loss or organ damage. Particular caution should be observed during surgery and with acute injuries.
Immunodeficiency	Hypogammaglobulinemi Immundefekt AIDS HIV	D80.0 Hereditary hypogammaglobulinaemia D84 D81 D82.0 B23	Patients often have serious infections masked as ordinary infections. Require special vigilance and prevention of infection following surgical procedures.
Isovaleric acidemia		E71.3 Other disorders in fatty acid metabolism	Metabolic disease where the patient often follows a low-protein diet with amino acid supplements. The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral nutrition.
Jervell-Lange-Nielsen			A congenital conduction disturbance in the heart causing the patient to be at risk of life-threatening arrhythmia. Patients must avoid a number of medicinal products that prolong the QT-interval.
Kallmann syndrome			Can result in hypopituitarism that leads to failure in the production of one or more hormones. The symptoms can vary greatly, including degrees of adrenal failure, hypothyroidism, diabetes insipidus and more. One must be aware of electrolyte disturbances and the need for hormone substitution.

Long-QT syndrome	Forlenget QT-tid Forlenget QT-tid-syndrom		A congenital conduction disturbance in the heart causing the patient to be at risk of life-threatening arrhythmia. Patients must avoid a number of medicinal products that prolong the QT-interval.
LCHAD			Metabolic disease The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral glucose polymer or parenteral nutrition containing 12-15% glucose via the nasogastric route. Lipids must not be administered intravenously.
Loeys-Dietz syndrome			Congenital connective tissue disease with significant risk of dilatation of- or acute bleeding from major arteries
Lorain-Levi syndrome			Hypopituitarism that leads to failure in the production of one or more hormones. The symptoms can vary greatly, including degrees of adrenal failure, hypothyroidism, diabetes insipidus and more. One must be aware of electrolyte disturbances and the need for hormone substitution.
Malignant brain tumour	Hjernesvulst Tumor cerebri Neoplasma malignum cerebri Ondartet svulst i hjerne Ondartet svulst i CNS	C71 Malignant neoplasm of brain (neoplasm malignum cerebri) C71.1 frontal lobe (Lobus frontalis) C71.2 temporal lobe (Lobus temporalis) C71.3 parietal lobe (Lobus parietalis) C71.4 occipital lobe (Lobus occipital) C71.5 Cerebral ventricle C71.6 Cerebellum C71.7 brainstem	Brain tumours can cause a number of complications, both in the form of bleeding, epileptic seizure etc.

		C71.8 Overlapping tumour of brain C71.9 Brain, unspecified	
Malignant hyperthermia	Ondartet hypertermi Hypertermi under anestesi	T88.3 Malignant hyperthermia due to anesthesia	Malignant hyperthermia is a sudden reaction to certain anaesthetics that can be life-threatening. Individuals who have experienced malignant hyperthermia must avoid triggering agents in the future.
Maple syrup urine disease	MSUD Amino acid disorders Aminosyre	E71.0 «Maple syrup urine disease»	Metabolic disease where the patient often follows a low-protein diet with amino acid supplements. The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral nutrition.
Marfan syndrome	Morbus Marfan Marfans sykdom Mb Marfan Mb. Marfan	Q87.4 Marfan syndrome	Congenital connective tissue disease, including changes in the cardiovascular system. Increased risk of dilation and rupture of the aorta and detached lenses
Maroteaux-Lamys disease (MPS VI)			Mucopolysaccharidoses are hereditary, congenital metabolic diseases caused by defective degradation of mucopolysaccharides. Acute situation/anesthesia: Atlantoaxial instability occurs. Risk of crushing the medulla.

Mastocytosis	Urticaria pigmentosa	Q82.2 Mastocytosis	Disease due to collection of excess mast cells, i.e. cells important in allergic reactions. Patients with mastocytosis may experience a powerful allergy-like reaction, which can lead to rapid onset of shock when they are exposed to: wasp stings, opiates, contrast media, NSAID or other medicines, foodstuffs, all types of food, latex, physical stress, heat, cold etc.). This must be treated as anaphylaxis/anaphylactic shock, depending on the severity. Patients should have an "EpiPen".
MCAD			Metabolic disease The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral glucose polymer or parenteral nutrition containing 12-15% glucose via the nasogastric route. Lipids must not be administered intravenously.
MELAS	Mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes Mitokondriesykdom Mitokondriemyopati Mitokondriopati G31.8 Other specified degenerative diseases of the central nervous system G71.3 Mitochondrial myopathy, not elsewhere classified		MELAS is a hereditary mitochondrial disease that attacks the central nervous system. The patient may suffer stroke-like episodes, which can be misinterpreted as epilepsy. Other key organs such as the heart and kidneys may also be affected.

Methylmalonic acidemia	METHYLMALONIC ACIDEMIA MMA Organic acidemias		Metabolic disease where the patient often follows a low-protein diet with amino acid supplements. The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral nutrition.
Mitochondrial Disease	Mitokondriesykdom Mitokondriemyopati Mitokondriopati Kearns-Sayre MNGIE	G71.3 Mitochondrial myopathy, not classified elsewhere	Mitochondriopathies require special measures during treatment, e.g. in infections/sepsis. Among other things, aminoglycosides and mecillinam are strongly contraindicated in certain mitochondriopathies. Sufferers can also become seriously ill following short periods without food, e.g. stomach upset.
Morquios syndrome (MPS IV)			Mucopolysaccharidoses are hereditary, congenital metabolic diseases caused by defective degradation of mucopolysaccharides. Acute situation/anesthesia: Atlantoaxial instability occurs. Risk of crushing the medulla.
Moyamoya disease		I67.5 Moyamoya disease	Moyamoya disease is caused by blocked arteries at the base of the brain. May cause stroke with diffuse symptoms.
MPS IX			Mucopolysaccharidoses are hereditary, congenital metabolic diseases caused by defective degradation of mucopolysaccharides. Acute situation/anesthesia: Atlantoaxial instability occurs. Risk of crushing the medulla.
Mucopolysaccharidosis	Mukopolysakkarid-ose MPS Hurlers sykdom (MPS I) Hunters sykdom (MPS II)	E76 Disorders of glucosaminoglycan metabolism	Mucopolysaccharidoses are hereditary, congenital metabolic diseases caused by defective degradation of mucopolysaccharides. Acute situation/anesthesia:

	Sanfilippos sykdom (MPS III) Morquios sykdom (MPS IV) Maroteaux-Lamys sykdom (MPS VI) Slys sykdom (MPS VII) MPS IX		Atlantoaxial instability occurs. Risk of crushing the medulla.
Muscular dystrophie Becker			Muscular diseases can affect respiration after anesthesia. May also have undiagnosed cardiomyopathy or conduction disturbances. Requires individual investigation prior to anesthesia. Increased effects of muscle relaxants than others, and also increased risk of developing malignant hyperthermia requires special choice of anesthetic agents.
Muscular dystrophie Duchenne			Muscular diseases can affect respiration after anesthesia. May also have undiagnosed cardiomyopathy or conduction disturbances. Requires individual investigation prior to anesthesia. Increased effects of muscle relaxants than others, and also increased risk of developing malignant hyperthermia requires special choice of anesthetic agents.
Muscular dystrophie Emery-Dreifuss			Muscular diseases can affect respiration after anesthesia. May also have undiagnosed cardiomyopathy or conduction disturbances. Requires individual investigation prior to anesthesia. Increased effects of muscle relaxants than others, and also increased risk of developing malignant hyperthermia requires special choice of anesthetic agents.
Muscular dystrophies/myopathies	Pompes sykdom Muskeldystrofi Emery-Dreifuss Muskeldystrofi Duchenne Muskeldystrofi Becker Dystrofia Myotonica	G71.0 Muscular dystrophy G71.1 Myotonia disorders G71.2 Congenital myopathies	Muscular diseases can affect respiration after anesthesia. May also have undiagnosed cardiomyopathy or conduction disturbances. Requires individual investigation prior to anesthesia. Increased effects of muscle relaxants than others,

	Steinerts sykdom Myotone lidelser Thomsens sykdom Erb-Duchenne palsy		and also increased risk of developing malignant hyperthermia requires special choice of anesthetic agents.
Myasthenia gravis	Myasteni	G70.0 Myasthenia gravis	An autoimmune disease of the nervous system which affects respiration. Up to one fifth of patients may experience crises where they develop acute respiratory failure. Requires caution during anesthesia.
Noonan syndrome			Noonan syndrome (NS) is a congenital disorder. The principal features are a characteristic configuration of facial features and include congenital heart defect, short stature, learning problems, pectus excavatum, and impaired blood clotting. May cause bleeding problems.
Organic aciduria	Glutarsyreuri Ornitintranscarbamylasemangel Organic acidemia	E72.3 Disorders in the metabolism of lysine and hydroxylysine E72.4 Disorders in ornithine metabolism	Metabolic disease The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral glucose polymer or parenteral nutrition containing 12-15% glucose via the nasogastric route.
Ornithine transcarbamylase (OTC) deficiency			Metabolic disease where the patient often follows a low-protein diet with amino acid supplements. The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral nutrition.

Osler's disease	Oslers sykdom Mb Osler Arvelig hemoragisk teleangiektasi Hereditær hemoragisk teleangiektasi HHT	I78.0 Hereditary haemorrhagic telangiectasis	Causes frequent nosebleeds and oozing haemorrhage in the gastrointestinal tract. May have vascular malformations in the lungs, brain and liver, which can cause shortness of breath and risk of bleeding and abscesses. Patients with untreated malformations in the lungs must have antibiotic prophylaxis on dental treatment and surgical procedures.
Osteogenesis imperfecta	OI	Q78.0 Osteogenesis imperfecta	Hereditary connective tissue disease, which includes increased risk of fracture. Many sub-groups. CAUTION: instability/limited extension of the neck, and special precautions during anesthesia. Careful handling to avoid fracture
Paraneoplastic syndrome	Paraneoplasi		A number of life-threatening situations may arise as a result of cytokine- or hormone production from a cancerous growth- or immune response to this.
Paroxysmal nocturnal haemoglobinuria	Marchiafava-Micheli syndrom	D59.5 Paroxysmal nocturnal haemoglobinuria	Rare disorder that causes the destruction of red blood cells. May cause leukopenia and thrombocytopenia. Requires caution during anesthesia and special prophylaxis against thrombosis.
Phenylketonuria	Føllings sykdom Mb. Følling PKU	E70.0 Classic phenylketonuria	A congenital disorder of the metabolism of the amino acid, phenylalanine. Patients need a special low-protein diet to preventserious symptoms.

Pheochromocytoma	Feocromocytom Phaeochromo-cytoma Binyremarghyper-plasi Hypersekresjon av katekolamin Adrenomedullary hyperfunction E27.5 D35.0		Pheochromocytoma is a hormone-producing tumour, usually benign. Caution must be observed during anesthesia and with certain drugs, since these may trigger complications.
Pituitary tumor			Can result in hypopituitarism that leads to failure in the production of one or more hormones. The symptoms can vary greatly, including degrees of adrenal failure, hypothyroidism, diabetes insipidus and more. One must be aware of electrolyte disturbances and the need for hormone substitution.
Pompe disease			Muscular diseases can affect respiration after anesthesia. May also have undiagnosed cardiomyopathy or conduction disturbances. Requires individual investigation prior to anesthesia. Increased effects of muscle relaxants than others, and also increased risk of developing malignant hyperthermia requires special choice of anesthetic agents.
Porphyria	Akutt porfyri AIP Akutt intermitterende porfyri E80.2 Annen porfyri Porfyria acuta Hereditær koproporfyri (HCP) Porphyria variegate (PV)	E80.0	Acute porphyry diseases can cause life-threatening acute neuro-visceral seizures that can be triggered by, inter alia, a number of common drugs. See www.napos.no for more information on which medicines should be avoided and information on the treatment of acute attacks.

Prader-Willi syndrome			Congenital condition characterized by varying degrees of developmental disability and / or learning disabilities. High pain threshold can lead to overlooked fractures and misdiagnosis of abdominal pain. Emergency medical consequence: Central apnea, decreased response to hypercapnia, narrow oropharyngeal conditions.
Previous splenectomy	Fjernet milt Postoperativ aspleni Aspleni Miltatrofi	D73.0 Hyposplenism	Patients having undergone splenectomy are at significantly higher risk of serious systemic pneumococcal disease, with extremely rapid development of illness and high mortality. Patients having undergone splenectomy should, therefore, be informed of this and followed up closely by their GP, both with a view to vaccination and early antibiotic treatment in the event of respiratory infections.
Prior subarachnoid hemorrhage	SAH Hjernehinneblødning Hemorrhagia subaracnoidalis	I60 Subarachnoid haemorrhage I60.0Subarachnoid haemorrhage from carotid siphon and bifurcation I60.1 Subarachnoid haemorrhage from middle cerebral artery I60.2 Subarachnoid haemorrhage from anterior communicating artery I60.3 Subarachnoid haemorrhage from posterior communicating artery I60.4 Subarachnoid haemorrhage from basilar artery I60.5 Subarachnoid haemorrhage from vertebral artery I60.6 Subarachnoid haemorrhage from other intracranial arteries I60.7 Subarachnoid haemorrhage from intracranial artery,	A type of bleeding in the brain with high chance of recurrence.

		unspecified I60.8 Other subarachnoid haemorrhage I60.9 Subarachnoid haemorrhage, unspecified	
Propionic acidemia	Amino acid disorders Organic acidemias Organisk aciduri		Metabolic disease where the patient often follows a low-protein diet with amino acid supplements. The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral nutrition.
Protein C deficiency	Trombofilia		Patients with Protein C deficiency have an abnormal tendency to form blood clots. Blood clots may stick in the lungs and brain and cause life-threatening complications. Diagnostic vigilance is required, and special precautions must be implemented in a variety of treatments.
Protein S deficiency	Trombofilia		Patients with Protein S deficiency have an abnormal tendency to form blood clots. Blood clots may stick in the lungs and brain and cause life-threatening complications. Diagnostic vigilance is required, and special precautions must be implemented in a variety of treatments.
Prothrombin mutation			Patients with mutation in the Protrombin gen have an abnormal tendency to form blood clots. Blood clots may stick in the lungs and brain and cause life-threatening complications. Diagnostic vigilance is required, and

			special precautions must be implemented in a variety of treatments.
Pseudocholinesterase deficiency	Cholinesterase-mangel Kolinesterasemangel Pseudokolinesterase mangel		Enzyme deficiency that causes muscle relaxants to degrade later than normal and may cause paralysis and respiratory failure for a long time after administration.
Pulmonary hypertension	Eisenmenger syndrom	I27.9 Pulmonary heart disease, unspecified I27.0 Primary pulmonary hypertension I27.2 Other secondary pulmonary hypertension	Eisenmenger syndrome is pulmonary hypertension caused by congenital malformations of the heart. The patients have heart failure, respiratory failure and may experience a tendency to both bleeding and clotting.
Romano-Ward syndrome			A disturbance in the heart causing the patient to be at risk of life-threatening arrhythmia. Patients must avoid a number of medicinal products that prolong the QT-interval.
Type II respiratory failure	KOLS Respiratory disorders Pulmonary disease	J96.1 Chronic respiratory failure	Type II respiratory failure is defined as hypoxia with hypercapnoea. Such patients have had prolonged exposure to high levels of CO2 in the blood and have developed tolerance to it. There is a risk that administration of oxygen to the patient will cause CO2-narcosis, since they no longer react to high CO2-concentrations in the blood.
Sanfilippo syndrome (MPS III)	Mucopoly-saccharidosis		Mucopolysaccharidoses are hereditary, congenital metabolic diseases caused by defective degradation of mucopolysaccharides. Acute situation/anesthesia: Atlantoaxial instability occurs. Risk of crushing the medulla.

Sarcoidosis	Sarcoidose Sarcoidosis	D86 Sarcoidosis D86.0 Sarcoidosis of lung D86.1 Sarcoidosis of lymph nodes D86.2 Sarcoidosis of lung with sarcoidosis of lymph nodes D86.3 Sarcoidosis of skin D86.8 Sarcoidosis of other and combined sites D86.9 Sarcoidosis, unspecified	May develop hyperkalaemia causing non-specific symptoms, with risk of cardiac rhythm disturbance.
Scott syndrom	Purpura/koagula-sjonsdefekt		Dysfunctional platelets results in delayed coagulation of the blood, such that the patient may die from blood loss or organ damage. Particular caution should be observed during surgery and with acute injuries.
Sheehan's syndrome			Syndrom that can result in hypopituitarism that leads to failure in the production of one or more hormones. The symptoms can vary greatly, including degrees of adrenal failure, hypothyroidism, diabetes insipidus and more. One must be aware of electrolyte disturbances and the need for hormone substitution.
Shwachman-Diamond syndrome (SDS)	Shwachman-Bodian-Diamond syndrom Shwachman-Diamond-Oski syndrome		Hereditary disorder with involvement of bone marrow, pancreas, liver and bone. Severe, acute bone marrow failure, myelodysplastic syndrome, severe life-threatening bacterial infections requiring intensive care, bleeding episodes
Simmonds' syndrom			Syndrom that can result in hypopituitarism that leads to failure in the production of one or more hormones. The symptoms can vary greatly, including degrees of adrenal failure, hypothyroidism, diabetes insipidus and more. One must be aware of electrolyte disturbances and the need for hormone substitution.

Situs inversus	Malrotasjon	Q89.3 Situs inversus	Patients have transverse localization of organs, which can cause significant diagnostic misunderstandings.
Slys' disease (MPS VII)			Mucopolysaccharidoses are hereditary, congenital metabolic diseases caused by defective degradation of mucopolysaccharides. Acute situation/anesthesia: Atlantoaxial instability occurs. Risk of crushing the medulla.
Spina bifida	Myelomeningocele Spina Bifida Meningomyelocele Syringomyelocele Arnold Chiari malformasjon	Q05 Spina bifida Q07.0 Arnold-Chiaris syndrom	Congenital neural tube defect. CAUTION: possible shunt failure and complications from possible Arnold Chiari malformation during anesthesia and surgery
Steinert's disease			Muscular diseases can affect respiration after anesthesia. May also have undiagnosed cardiomyopathy or conduction disturbances. Requires individual investigation prior to anesthesia. Increased effects of muscle relaxants than others, and also increased risk of developing malignant hyperthermia requires special choice of anesthetic agents.
Storage pool deficiencies	Purpura/koagulasjon sdefekt		Dysfunctional platelets results in delayed coagulation of the blood, such that the patient may die from blood loss or organ damage. Particular caution should be observed during surgery and with acute injuries.

Sturge Weber syndrome	encephalotrigeminal angiomatosis		Sturge Weber -patients have a tendency to experience stroke-like episodes, with subsequent hemiparesis. These may be difficult to distinguish from epileptic seizures, but need different strategies for assessment and treatment.
Thrombocytopenia	Purpura Blodplatedefekt Trombocytopenisk purpura ITP Idiopatisk trombocytopeni	D69.0 Allergic purpura D69.1 Qualitative platelet defects D69.2 Other non-thrombocytopenic purpura D69.3 Idiopathic thrombocytopenic purpura D69.4 Other primary thrombocytopenia D69.5 Secondary thrombocytopenia D69.6 Thrombocytopenia, unspecified	Lack of- or dysfunctional platelets results in delayed coagulation of the blood, such that the patient may die from blood loss or organ damage. Particular caution should be observed during surgery and with acute injuries.
Thrombophilia	Blodpropptendens Aktivert protein C resistens Faktor V Leidenmutasjon Antitrombinmangel Protein C-mangel Protein S-mangel Protrombingen-mutasjon Antikardiolipin-syndrom Antifosfolipid-syndrom Tilstedeværelse av lupus antikoagulant APC	D68.5 Primary thrombophilia D68.6 Other thrombophilia	Patients with thrombophilia have an abnormal tendency to form blood clots. Blood clots may stick in the lungs and brain and cause life-threatening complications. Diagnostic vigilance is required, and special precautions must be implemented in a variety of treatments.
Thomsen disease			Muscular diseases can affect respiration after anesthesia. May also have undiagnosed cardiomyopathy or conduction disturbances. Requires individual investigation prior to anesthesia. Increased effects of muscle relaxants than others, and also increased risk of developing malignant hyperthermia requires special choice of anesthetic agents.

Tuberous sclerosis	TSC Tuberous Sclerosis Complex Bourneville's disease	Q85.1 Tuberous sclerosis	Growth of benign tumors especially in brain, kidney, lungs and other organs. May cause seizures, and severe symptoms from affected organs.
Urea cycle defects	urea cycle disorder Defekt i urinsyresyklus Argininemi Arginino-suksininsyreuri Citrullinemi Hyperammonemi	E72.2 Disorders of urea cycle metabolism	Metabolic disease where the patient often follows a low-protein diet with amino acid supplements. The patient should avoid fasting. In conditions that lead to breakdown of the body's protein (injury, surgery, fever, vomiting, fasting etc.), acute treatment with an SOS-regime (glucose polymer) is required to avoid life-threatening metabolic decompensation. If vomiting is present, the patient must have parenteral nutrition.
Vascular malformations in the brain	Hjerneaneurisme Cerebrovaskulært aneurisme Cerebral aneurysm Moyamoyasykdom	I67.1 Cerebral aneurysm, non-ruptured Q28.2 Arteriovenous malformation in cerebral vessel Q28.3 Other malformations of cerebral vessels I67.5 Moyamoya disease	Malformations of cerebral vasculature, which can suddenly start to bleed. In some cases can cause diffuse symptoms, which do not lead one to suspect cerebral bleeding.
Wilson's disease	Hepatolentikulær degenerasjon Morbus Wilson		A genetic disorder in which copper builds up in the body. May cause liver failure,neuropsychiatric symptoms incl. epilepsy

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