

題	名 Title
第	ーに影響を受けるコード(または章番号)Primary Code Affected
第 [	ニに影響を受けるコード(オプション) Secondary Codes Affected
影	響を受ける巻(複数選択可)Volumes Affected
L	 Volume I (内容例示) Volume II (総論) Volume III (索引)
提	案の型 Proposal Type(どれか一つを選ぶ)
L	」 #1新しいコードの追加 Addition of new code
	#2コードの削除 Deletion of code
	#3コードの他カテゴリー(3桁分類項目または中間分類項目)または章への移動
	Movement of a code to another category or chapter
	#4新しい用語の索引への追加 Introduction of a new term into the index.
	#5既存の検索用語の割当コードの3桁分類項目レベルでの移動 Change to an
	existing index entry that changes the code assignment from one
	three-character category to another three-character category (movement of terms)
	#6既存の検索用語の割当コードの同じ3桁分類項目内での移動 Correction or
	clarification of existing index entry that only changes the code assignment to
	a code within the same three-character category
	#7内容例示の充実(包含、除外への追加など)Enhancements to the tabular list
	(such as the addition of an inclusion term to an existing code; the addition of an exclusion note)
	#8コード説明(注など)を疾病概念を変えない範囲で変更 Change to a code
	description that enhances the description rather than changes the concept
	・・・・・・・・・・・・・・・・・・・・・・・・・・・・・・・・・・・・
	ど) Enhancements to the index (such as the duplication of an existing index
	entry under another main term)

- #10疾病・死因データ収集規準に影響するルールまたはガイドラインの変更 Change to a rule or guideline that affects the integrity of morbidity or mortality data collections
- #11疾病・死因データ収集規準には影響しないルールまたはガイドラインの変更 Change to a rule or guideline that does not affect the integrity of morbidity or mortality data collections
- #12誤字・脱字等の修正 Correction of a typographical error
- #13出版物の修正には至らない明確化 Clarification that does not require change in the volumes

### 変更理由 Change Reason(どれか一つを選ぶ)

- #1新しい疾患を同定する必要があるため Need to identify a new disease.
- #2医療知識の変更を反映する必要があるため Need to reflect a change in clinical knowledge
- #3医療用語の変更を反映する必要があるため Need to reflect a change in clinical terminology
- #4他のファミリー分類との整合性を保つ必要があるから Need for compatibility within the WHO Family of Classifications
- #5ルール、ガイドラインまたは取り決めの変更による Change in rules, guidelines or conventions
- #6内容例示をより明確にするあるいは曖昧さをなくす必要があるから Need to improve clarity or reduce ambiguity in the tabular list
- #7検索用語を追加、削除または修正する必要があるから Need to create, delete or correct an index entry
- #8より詳細にあるいは反対に一般的にするなど詳細さのレベルを変える必要があるから Need for greater or less specificity
- #9誤字・脱字等の修正 Need to correct typographical errors

### 言語版に特徴的な改正 Update Specific to a Language Version (普通は該当しない)

### 詳細な記述 Detailed Description

(実際に出版物の記載がどう変更されるかを見え消しで記載する、提案の型で#13 を選んだとき以外は記載)

上	記の変更が必要なロジック Rationale
L	
上詞	記の変更を支持する論文等(もしあれば)Supporting Publication

(ファイル、URL など)

### **VATER** syndrome

Proposal ID: 1163 - Proposal State: In Open Discussion Layer

Proposal for Update

Originator: Ulrich Vogel - Last Update made by: Lori Moskal

Creation Date: 22-Mar-2007 12:06 CET - Last Update:

30-Nov-2007 18:56 CET

**Previously Discussed in the group(s):** 

Primary Code Affected: Q87.2

**Secondary Codes Affected:** Q87.8

Volumes Affected: 1,3

**Proposal Type:** Enhancements to the tabular list (such as the addition of an inclusion term to an existing code; the addition of an exclusion note)

Change Reason: Need to reflect a change in clinical terminology

#### **Detailed Description**

Delete inclusion:

# Q87.2 Congenital malformation syndromes predominantly involving limbs

Syndrome:

- · Holt-Oram
- · Klippel-Trénaunay-Weber
- · nail patella
- · Rubinstein-Taybi
- · sirenomelia
- · thrombocytopenia with absent radius [TAR]

· VATER

Add inclusion:

Q87.8 Other specified congenital malformation syndromes, not elsewhere classified

Syndrome:

- · Alport
- · Laurence-Moon(-Bardet)-Biedl
- · Zellweger

**VATER** association

Revise code:

VATER association (syndrome) Q87.28

#### **Rationale**

The VATER syndrome is known today as VATER association which means that the linkage between the different malformations is not as strong as it is in syndromes (see ref.).

Apart from this in patients with VATER association type of malformation involvement of limbs is not predominantly (see ref.).

Thus we would suggest moving VATER from Q87.2 to Q87.8 and adding 'association'.

### **Supporting Publication Web Links**

VATER association at OrphaNet



#### **Hide Details**

Year/Round	Status	Your Vote	Results	
2008- 1	Voting process has not		Yes	0
	started for this round.		No	0
	Start Date :01/01/2008		Can't Dec	ide 0
			Not Voted	14

Year/Round	Status	Your Vote	Results

Year/Round	Status	Your Vote	Results	
2008- 2	Voting process has not		Yes	0
	started for this round.		No	0
	Start Date :02/06/2008		Can't Decide	0
			Not Voted	14

Year/Round	Status	Your Vote	Results	Results	
2008- 3	Voting process has not		Yes	0	
	started for this round.		No	0	
	Start Date :02/09/2008		Can't Dec	cide 0	
			Not Vote	d 14	

### **Comments**

Add a new comment

28-Jun-2007 02:42 CET by **Julie Rust** 

# Comment attached to the vote of the user for Round 1 of year 2007. Voted:Can't Decide

Agree with adding 'association' but not sure about moving to Q87.8. Also suggest adding the synonym 'VACTERL association' as an inclusion term and index entry.

06-Aug-2007 15:50 CET by Marion Mendelsohn

# Comment attached to the vote of the user for Round 2 of year 2007. Voted:Can't Decide

we agree with Julie's comment

30-Aug-2007 22:01 CET by **Robert Jakob** 

## Comment attached to the vote of the user for Round 2 of year 2007. Voted:Can't Decide

agree with code change, name edit and VACTERL, unless ORPHANET objects regarding the non-predominance of the limbs.

07-Sep-2007 19:04 CET by **Lori Moskal** 

# Comment attached to the vote of the user for Round 2 of year 2007. Voted:Can't Decide

AGree with Julie's comments. The following excerpt is from Oski's Pediatrics: The nonrandom association of vertebral defects, anal defects, tracheoesophageal fistula, and renal and radial abnormalities was initially designated as VATER association, but has been expanded to VACTERL to recognize the increased incidence of cardiac and limb defects. These patients usually do not show facial dysmorphism. The presence of at least three of the features is considered necessary for diagnosis.

18-Dec-2007 15:52 CET by Michael Schopen

## Comment attached to the vote of the user for Round 3 of year 2007. Voted:Can't Decide

Trieste decision was to hold over. Thus no voting.

