Checklist LQT (2011)

Findings			Points
ECG[1]	QTc[2]	≥ 480 ms	3
		= 460-479 ms	2
		= 450-459 ms (in males)	1
		≥ 480 ms during 4th minute of recovery from exercise stress test	1
	Torsade de pointes[3]		2
	T wave alternans		1
	Notched T wave in 3 leads		1
	Low heart rate for age[4]		0.5
Clinical history	**Syncope**[3]	**With stress**	2
		Without stress	1
Family history	Family member(s) with definite LQTS[5]		1
	Unexplained sudden cardiac death at age <30 years in immediate family[5]		0.5
Total score			X

[1]In the absence of medications or disorders known to affect these electrocardiographic features

[2]QTc (corrected QT) calculated by Bazett's formula where QTc = QT/√RR

[3]Mutually exclusive

[4]Resting heart rate <2nd %ile for age

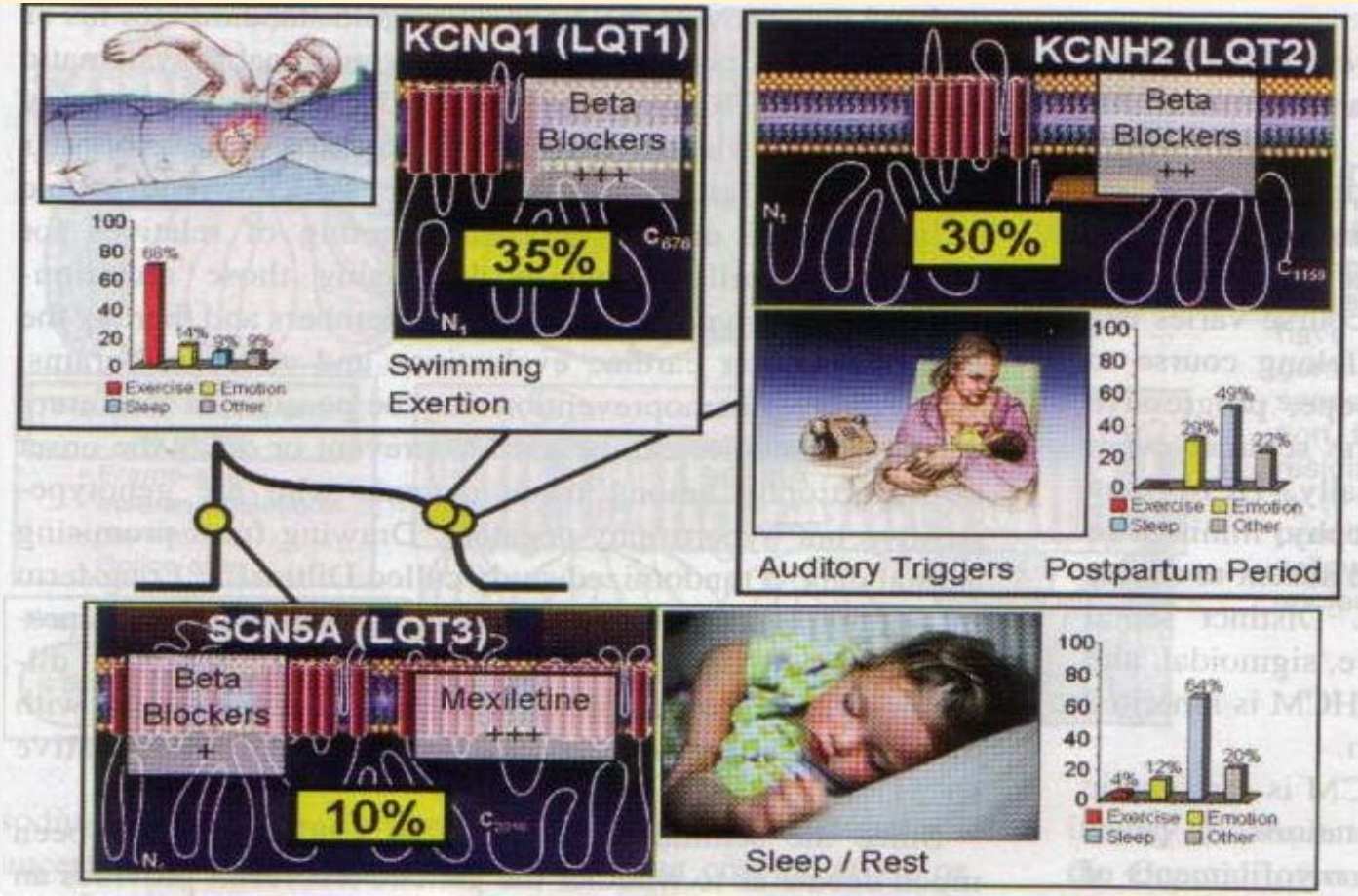
[5]The same family member cannot be counted for both criteria.

1. Schwartz PJ, Crotti L. QTc behavior during exercise and genetic testing for the long-QT syndrome. Circulation. 2011;124(20):2181-84.

Phenotypes and Genotypes in LQT – Syndrom (3 examples)

	LQT 1	LQT 2	LQT 3
Trigger	physical activity diving,swimming	emotional stress loud noises	Sleep low heart rate
First Mani- festation	< 9ys (53%) < 20 ys. (86 %)	- asymptomatic - < 16 ys.	(50%) - -
Sex Difference	in childhood	more in boys	later on more females
ß-Blocker	symptoms less (81%)	symptoms less (59%)	symptoms less (50%) first line:Mexiletin
Special:	cardiac events (63%)	Risk increased during pregnancy	events in 18%, but deadly in 64 %

Long QT-Syndrome: ECG ,Subgroups, and Clinical Aspects (Tester and Ackerman, Circulation 2011)



LQT-Syndrom : Score to Diagnosis

(Priori , 2015)

Score to diagnosis of LQT-Syndrom	
EKG - findings (no drugs or other known causes)	
QTc ≥ 480 ms (Bazzet-Formula)	3 Pts
QTc 460–479 ms	2 Pkt.
QTc 450–459 (m)	1 Pkt.
QTc > 480 ms (4 min after exercise)	1 Pkt.
Torsade de pointes without syncope	2 Pkt.
T- wave -alternans	1 Pkt.
T- wave - notch in 3 leads	1 Pkt.
Lower heart rate (below 2nd Percentil, age correctesde)	0,5 Pkt.
History	
Synkope, stress induced	2 Pkt.
Synkope,	1 Pkt.
Labyrinthine deafness, congenital	0,5 Pkt.
Family history	
LQTS in one member of family	1 Pkt.
Unexplained sudden cardia death <30 LJ	0,5 Pkt.

> 4 points : high; < 1 point: low probability,

Short QT-Syndrom or Early Repolarisation
(J-wave syndrom)
Def. : SQT + arrhythmia, rare syndrome (Herg gene)

Parameter		Punktwert
QTc (ms)	<370	1
	<350	2
	<330	3
J-Punkt bis zur Spitze von T (ms)	<120	1
Anamnese	Überlebter plötzlicher Herztod	2
	Polymorphe Kammertachykardie oder Kammerflimmern	2
	Unklare Synkope	1
	Vorhofflimmern	1
Familienanamnese	Angehöriger 1. oder 2. Grades mit SQTS	2
	Plötzlich verstorbener Angehöriger 1. oder 2. Grades	1
	Plötzlicher Kindstod	1
Genotyp	Positiver Genotyp	2
	Mutation von unbestimmer Bedeutung	1

In athletes mostly normal finding

Antzelevitch, J Arrhythm, 2016

Drug induced or aquired long-QT syndrom

El-Sherif,N. et al.Electrophys.Abl.(AER)2019,,8:122

Tabelle 2

Medikamenteninduzierte Verlängerung des QT-Intervalls und Torsade de pointes

Freiname	Handelsname	QT-Intervall	Torsade de pointes
Antiarrhythmika			
Ajmalin	Gilurytmal	+	+
Amiodaron	Cordarex, Cordarone	+	+
Chinidin	Cordichin (+ Verapamil)	+	+
Disopyramid	Norpace	+	+
Dofetilid	n. v.	+	+
Ibutilide	n. v.	+	+
Propafenon	Rytmonorm	+	+
Sotalol	Sotalex, Darob, Sotahexal	+	+
Antibiotika (Makrolide)			
Azithromycin	z. B. Zithromax		+
Clarythromycin	Klacid	+	+
Clindamycin	z. B. Sobelin, Clindahexal		+
Erythromycin	z. B. Eryhexal, Erycinum	+	+
Roxithromycin	z. B. Rulid	+	
Spiramycin	Rovamycine, Selectomycin	+	+
Antibiotika (Fluorchinolone)			
Gatifloxacin	Tequin	+	+
Grepafloxacin*1	Vaxar	+	+
Levofloxacin	Tavanic		+
Moxifloxacin	Avalox	+	+
Sparfloxacin	Zagam	+	+
Andere Antibiotika			
Ampicillin	z. B. Binotal	+	
Trimethoprim-Sulfamethoxazol	z. B. Bactrim	+	+
Antihistaminika			
Astemizol*1	Hismanal	+	+
Clemastin	Tavegil	+	
Diphenhydramin	z. B. Benadryl, Dolestan, Emesan	+	
Hydroxyzin	z. B. Atarax	+	
Terfenadin	z. B. Teldane	+	+
Antidepressiva			
Amitryptilin	z. B. Saroten	+	+
Clomipramin	Anafranil	+	
Desipramin	Pertofran, Pertylyl	+	+
Doxepin	z. B. Aponal	+	
Imipramin	z. B. Tofranil	+	+
Maprotilin	z. B. Ludiomil	+	+
Neuroleptika			
Amisulprid	Solian	+	
Clozapin	z. B. Leponex	+	
Chlorpromazin	Propaphenin	+	+
Droperidol*1	Dehydrobenzperidol	+	+
Fluphenazin	z. B. Dapotum, Lyogen	+	
Haloperidol	z. B. Haldol	+	+
Melperon	z. B. Eunerpan, Harmosin	+	+
Olanzapin	Zyprexa	+	
Pimozid	Orap	+	+
Quetiapine	Seroquel	+	
Sulpirid	z. B. Arminol, Dogmatil	+	+
Thioridazin	z. B. Melleril	+	+
Risperidon	Risperdal	+	
Sertindol*2	Serdolect	+	+
Tiaprid	Tiapridex	+	+
Trazodon	Thromban	+	
Selektive Serotonin-Wiederaufnahmehemmer			
Fluoxetin	z. B. Fluctin	+	+
Paroxetin	z. B. Seroxat, Tagonis	+	
Sertralin	Gladem, Zoloft	+	+
Antikonvulsiva			
Valproinat	z. B. Olfiril, Ergenyl	+	
Andere Psychopharmaka			
Chloralhydrat	Chloraldurat	+	+
Levomethadon	Orlaam	+	+
Lithium	z. B. Hypnorex, Quilonum	+	
Naratriptan	Naramig	+	
Sumatriptan	z. B. Imigran	+	
Venlafaxin	Trevilor	+	
Zolmitriptan	AscoTop	+	
Antiparkinsonmittel			
Amantadin	z. B. PK-Merz		+
Budipin*3	Parkinsan	+	+
Malariamittel			
Chinin	z. B. Limptar	+	+
Chloroquin	z. B. Resochin, Weimerquin	+	+
Halofantrin	Halfan	+	+
Mefloquin	Lariam	+	
Diuretika			
Indapamid	z. B. Natrilix, Sicco	+	
Lipidsenker			
Probucol	n. v.	+	+
Motalitätsanreger			
Cisaprid*1	Propulsin	+	+
Nootrope Geriatrika			
Vincamin	z. B. Ophdilvas	+	+
Chemotherapeutika			
Tamoxifen	z. B. Jenoxifen, Kessar, Nolvadex, Nourytam, Zemide	+	+
Pentamidin (i. v.)	Pentacarinat	+	+
Immunsuppressiva			
Tacrolismus	Prograf	+	+
Peptide			
Octreotid	Sandostatin	+	
Virostatikum			
Foscarnet	Foscavir	+	
Muskelrelaxans			
Tizanidin	Sirdalud	+	
Röntgen-Kontrastmittel			
Ioxaglinsäure	Hexabrix	+	+

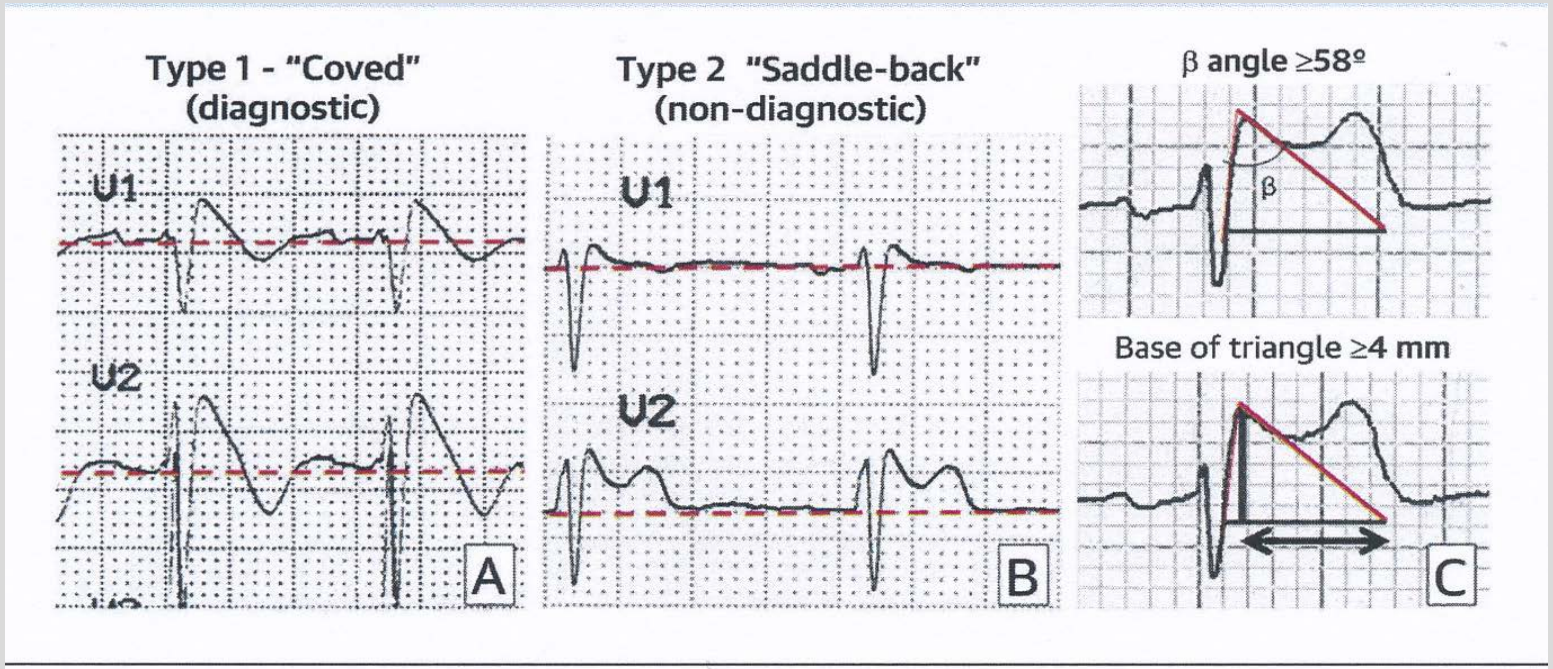
+ eine QT-Verlängerung kann auftreten bzw. Torsade de pointes wurden beobachtet; n. v., nicht in Deutschland im Handel verfügbar; *1 vom Markt zurückgenommen; *2 vom Markt suspendiert, endgültige Entscheidung durch die Zulassungsbehörden steht aus; *3 Anwendungsbeschränkungen ausgesprochen

Wichtige Hinweise zum Gebrauch der Tabelle: Die Tabelle stellt keinen Anspruch auf Vollständigkeit. Die Angaben beruhen auf dem aktuellen wissenschaftlichen Erkenntnisstand, soweit er öffentlich zugänglich ist (publizierte Studien (Medline-Recherchen), Fallberichte, Internet-Veröffentlichungen, Fachinformationen, Rote Liste, Mitteilungen von Zulassungsbehörden. In den verfügbaren Fallberichten über Torsade de pointes ist der kausale Zusammenhang mit der Einnahme der jeweiligen Substanz nicht mehr evident, eine reine Koinzidenz kann im Einzelfall nicht ausgeschlossen werden.

Check for drugs:
e.g. antihistaminics
(e.g. Terfenadin) plus
Makrolides - Antibiotics

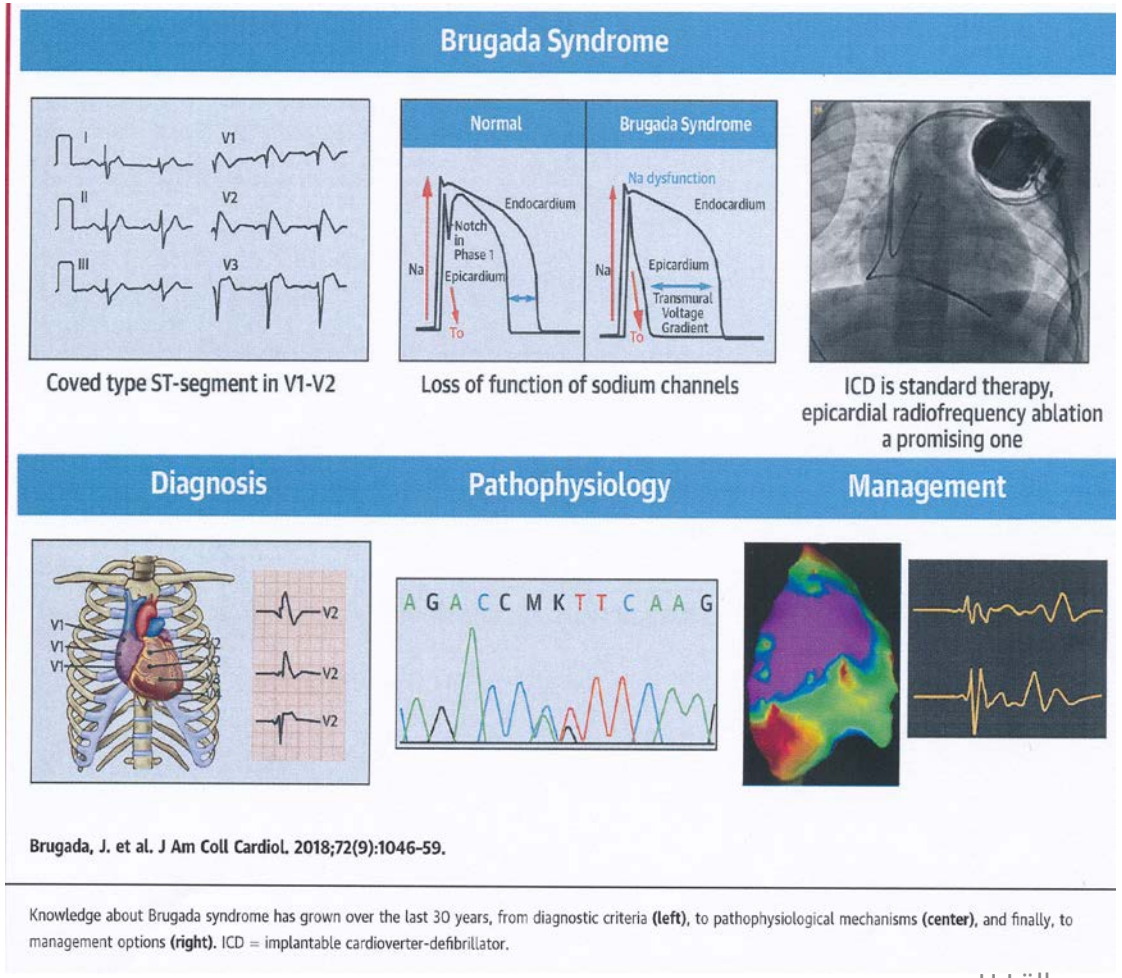
www.qt-syndrome.org.

Brugada Syndrom



rugada electrocardiogram pattern showing a concave ST-segment elevation ≥2 mm in ≥1 right precordial lead, foll
wave. **(B)** Type 2 Brugada electrocardiogram pattern showing a convex ST-segment elevation ≥0.5 mm (generally ≥2
dial lead followed by a positive T-wave. **(C)** Additional criteria for the diagnosis of Brugada electrocardiogram pattern
described by Chevallier et al. (18); **bottom:** the length of the base triangle of the r′ wave 5 mm below the maximum r

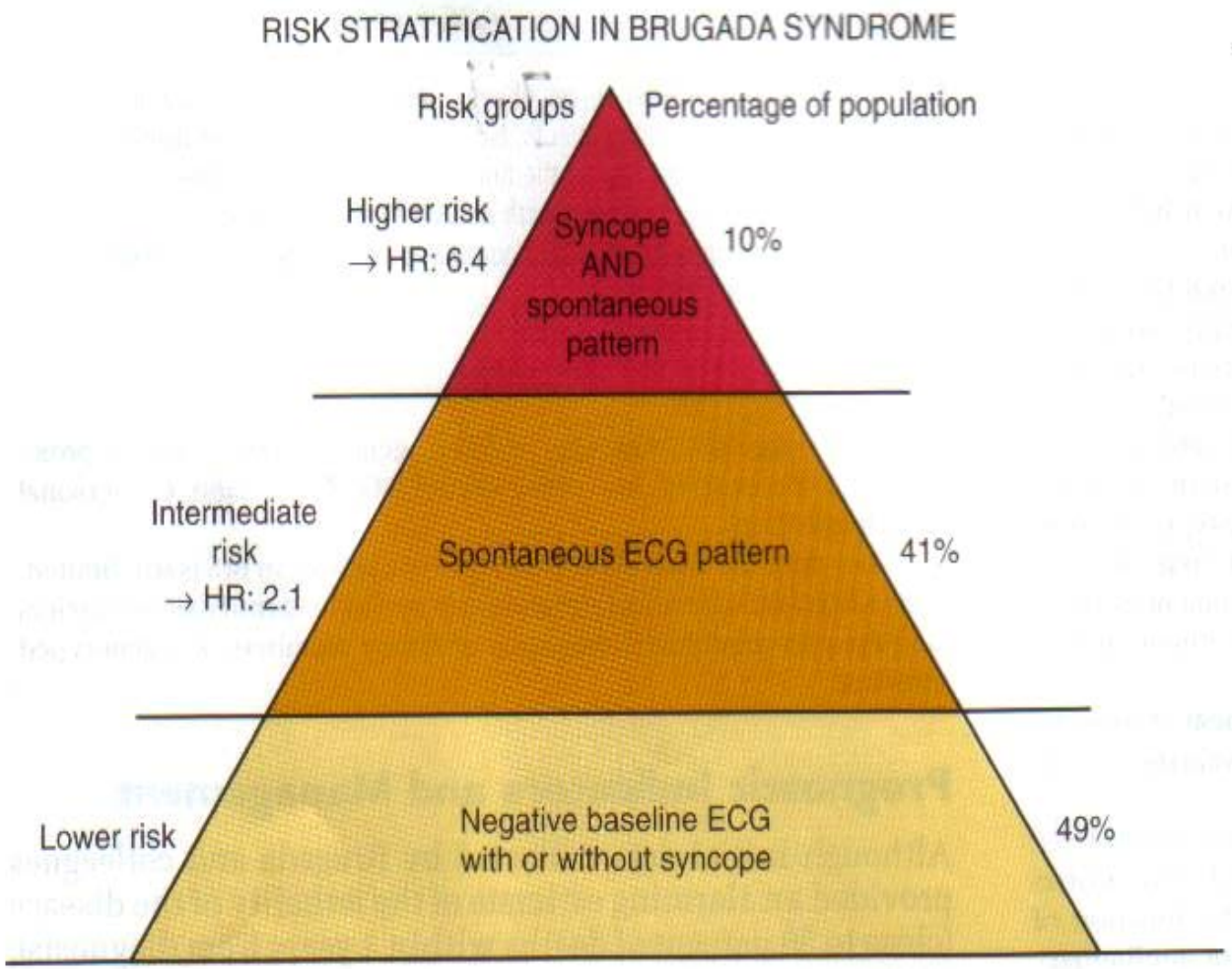
Brugada Syndrome: RBBB saddle or coved form, ST elevation in V1, V2, more pronounced after Ajmalin or Flecainid (Brugada et al., JACC 2018)



	Type 1	Type 2	Type 3
J point	> 2 mm	>2 mm	> 2mm
ST	Coved	saddle	
Isoelectr	--	> 1mm	<1mm
T wave:	neg	pos	pos

Risk Stratification in Brugada-Syndrom

(Priori,2008)



Cardiac arrest: 11%,

Synkope in 17 % of 200 pts.

EPS ?

Genetic Examination,

In pt. and in the family

Catecholaminergic Polymorph Ventricular Tachycardia (CPVT) (rare) (Coumel,1978)

- **Genetics:**
- autosomal dominant or recessiv, (Gen : RyR2 (Priori, 2001))
- Induced by stress or vigorous exercise
- **ECG** : normal (Idiopatic VF)
- Arrhythmia : VT (bidirectional or biventricular), V fib possible
  supraventrikular T.,AFib,
- Therapy : ß-Blocker or ICD
- **Diagnosis: Exercise -ECG** up to real exhaustion and/or exercising
  at 120 – 140 bpm physician in the room is mandatory

Inherited Progressive Conduction Disturbances
(e.g. Lev-Leneggre-Symptome)

- AV-Block I – III
- RBBB
- QRS-wave prolongation with /without PQ-prolongation

Priori et al. Ex.Summary HRS/EHTRA/APHRS expert consensus statement, Europace 2013, ESC GL 2015

Marfan Syndrom: The Revised Ghent Nosology

B.L Loeys et al.J.Med.Genet.2010:476

In the absence of family history:
(1) Ao (Z ≥2) AND EL=MFS*
(2) Ao (Z ≥2) AND *FBN1*=MFS
(3) Ao (Z ≥2) AND Syst (≥7pts)=MFS*
(4) EL AND *FBN1* with known Ao=MFS

EL with or without Syst AND with an *FBN1* not known with Ao or no *FBN1*=ELS
Ao (Z < 2) AND Syst (≥5 with at least one skeletal feature) without EL=MASS
MVP AND Ao (Z <2) AND Syst (<5) without EL=MVPS

In the presence of family history:
(5) EL AND FH of MFS (as defined above)=MFS
(6) Syst (≥7 pts) AND FH of MFS (as defined above)=MFS*
(7) Ao (Z≥2 above 20 years old, ≥3 below 20 years) +FH of MFS (as defined above)=MFS*

* Caveat: without discriminating features of SGS, LDS or vEDS (as defined in table 1) AND after *TGFBR1/2*, collagen biochemistry, *COL3A1* testing if indicated. Other conditions/genes will emerge with time.
Ao, aortic diameter at the sinuses of Valsalva above indicated Z-score or aortic root dissection; EL, ectopia lentis; ELS, ectopia lentis syndrome; *FBN1*, fibrillin-1 mutation (as defined in box 3); *FBN1* not known with Ao, *FBN1* mutation that has not previously been associated aortic root aneurysm/dissection; *FBN1* with known Ao, *FBN1* mutation that has been identified in an individual with aortic aneurysm; MASS, myopia, mitral valve prolapse, borderline (Z<2) aortic root dilatation, striae, skeletal findings phenotype; MFS, Marfan syndrome; MVPS, mitral valve prolapse syndrome; Syst, systemic score (see box 2); and Z, Z-score.

Box 2 Scoring of systemic features

- Wrist AND thumb sign — 3 (wrist OR thumb sign — 1)
- Pectus carinatum deformity — 2 (pectus excavatum or chest asymmetry — 1)
- Hindfoot deformity — 2 (plain pes planus — 1)
- Pneumothorax — 2
- Dural ectasia — 2
- Protrusio acetabuli — 2
- Reduced US/LS AND increased arm/height AND no severe scoliosis — 1
- Scoliosis or thoracolumbar kyphosis — 1
- Reduced elbow extension — 1
- Facial features (3/5) — 1 (dolichocephaly, enophthalmos, downslanting palpebral fissures, malar hypoplasia, retrognathia)
- Skin striae — 1
- Myopia > 3 diopters - 1
- Mitral valve prolapse (all types) — 1

Maximum total: 20 points; score ≥7 indicates systemic involvement; US/LS, upper segment/lower segment ratio.

Conclusion I
Role of Checklists

- Occult or latent diseases in cardiology are rare, but possible fatal even as first manifestation

- To recognize these inherited diseases,
- checklists are helpful for the less experienced physician as well for the cardiologist

- These diseases occur in children and younger persons during exercise and swimming (36%) and (DD. accident)

Conclusion II
Role of Checklists

If occult diseases are present, they may

- **do harm to the heart,**
- **cause severe arrhythmias**
- **cause sudden cardiac arrest**

Therefore, checklists may support the physician, esp. together with a special software support by ECG device

ECG 2017: International Criteria