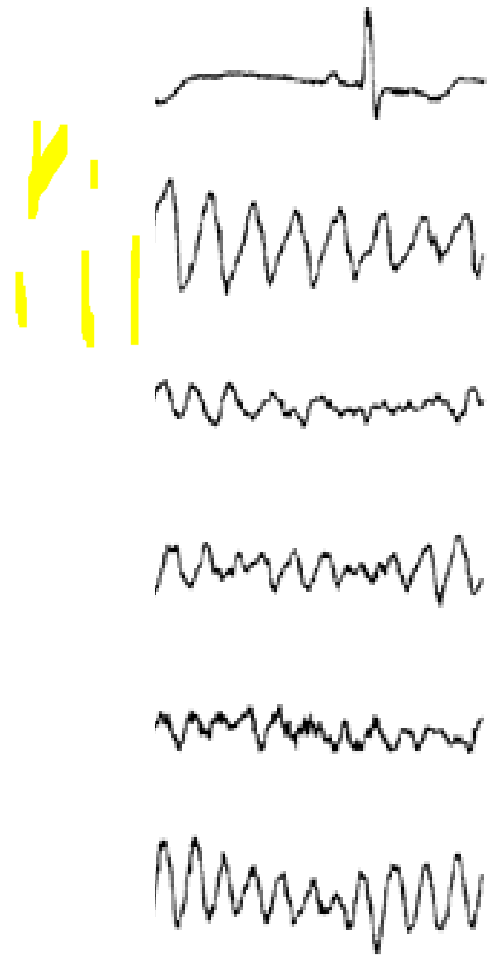
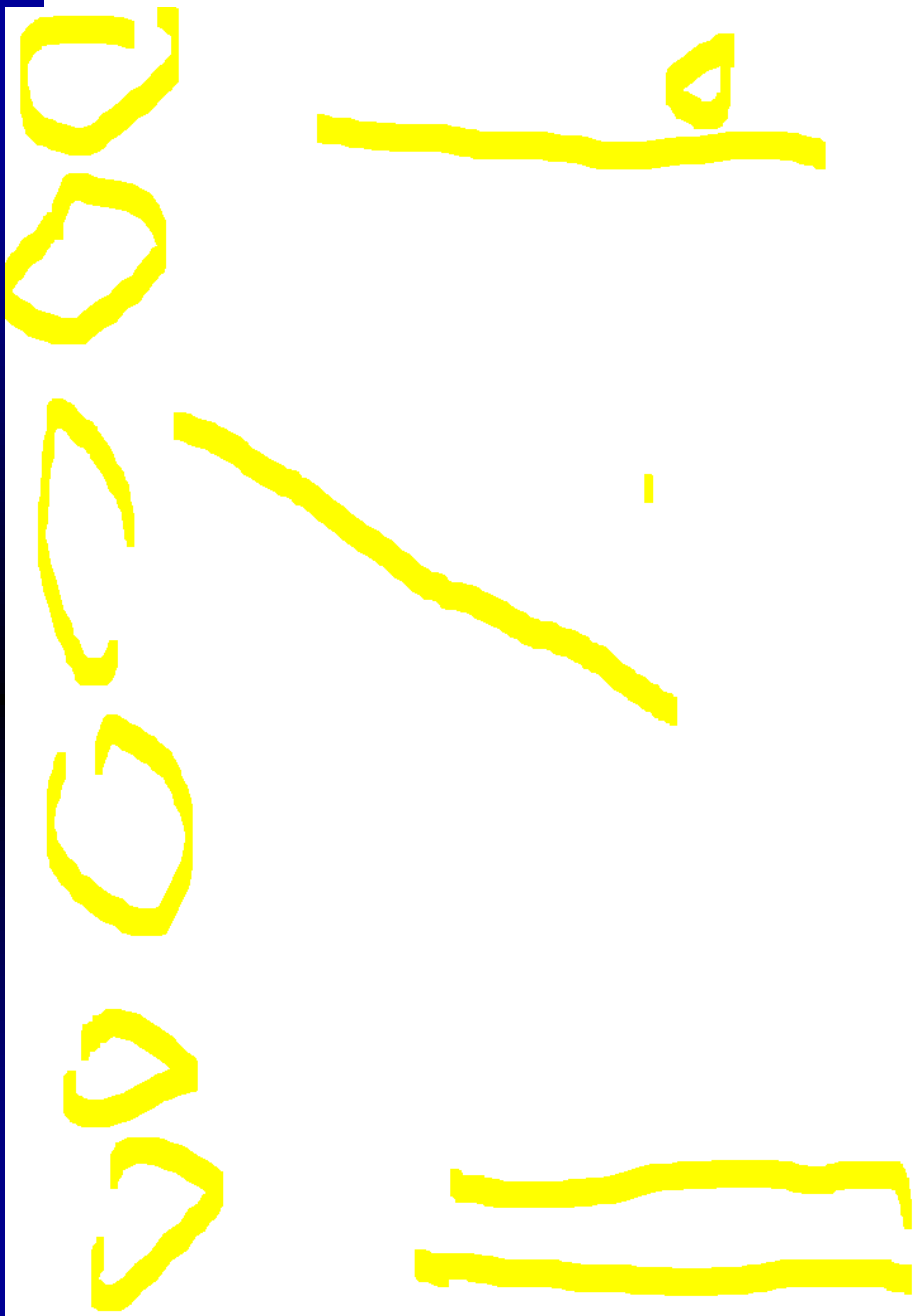


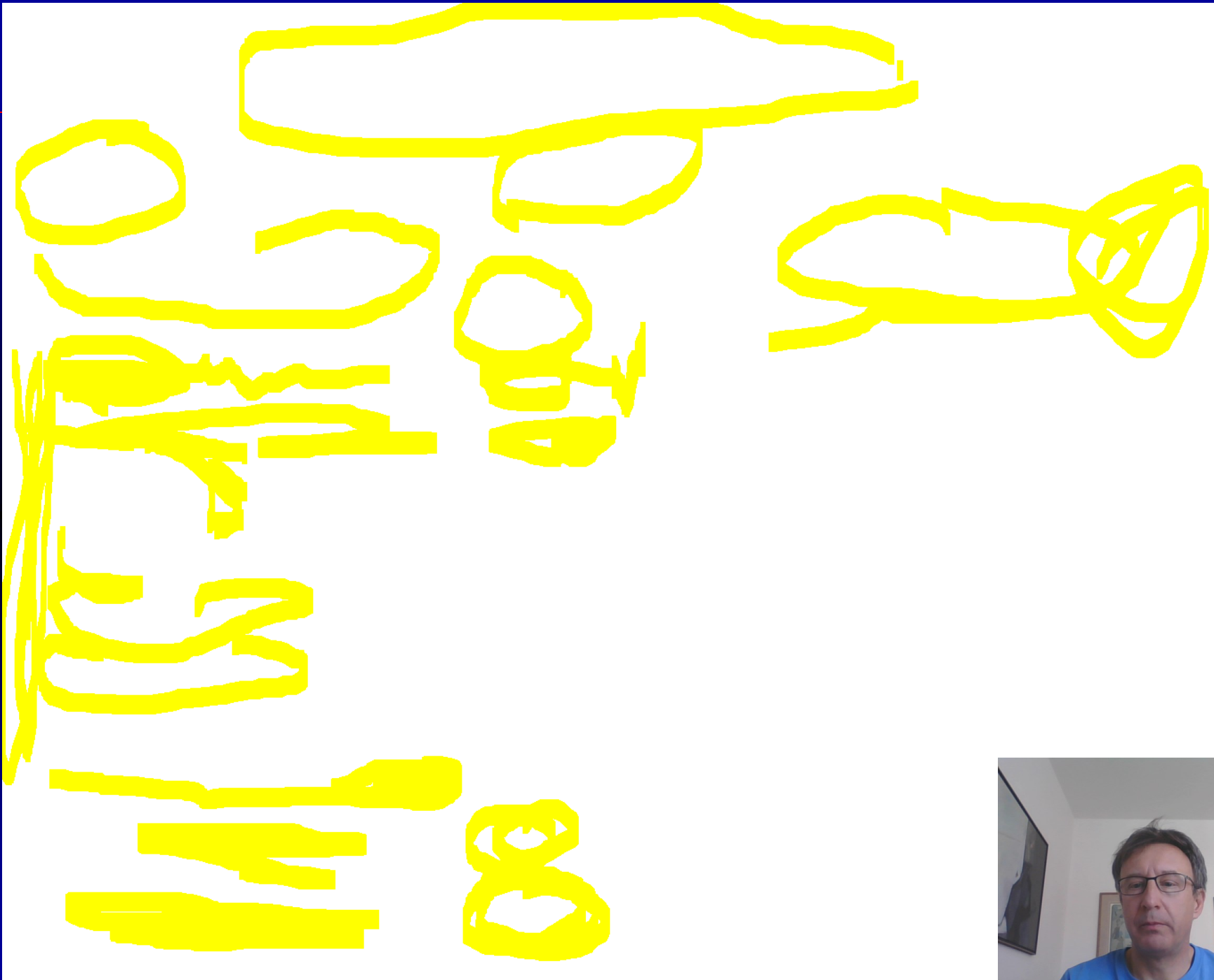


J. G. Mendel





(D)





Pathophysiology of SCD on molecular level

1) processes of electric impulse creation and propagation in myocardium

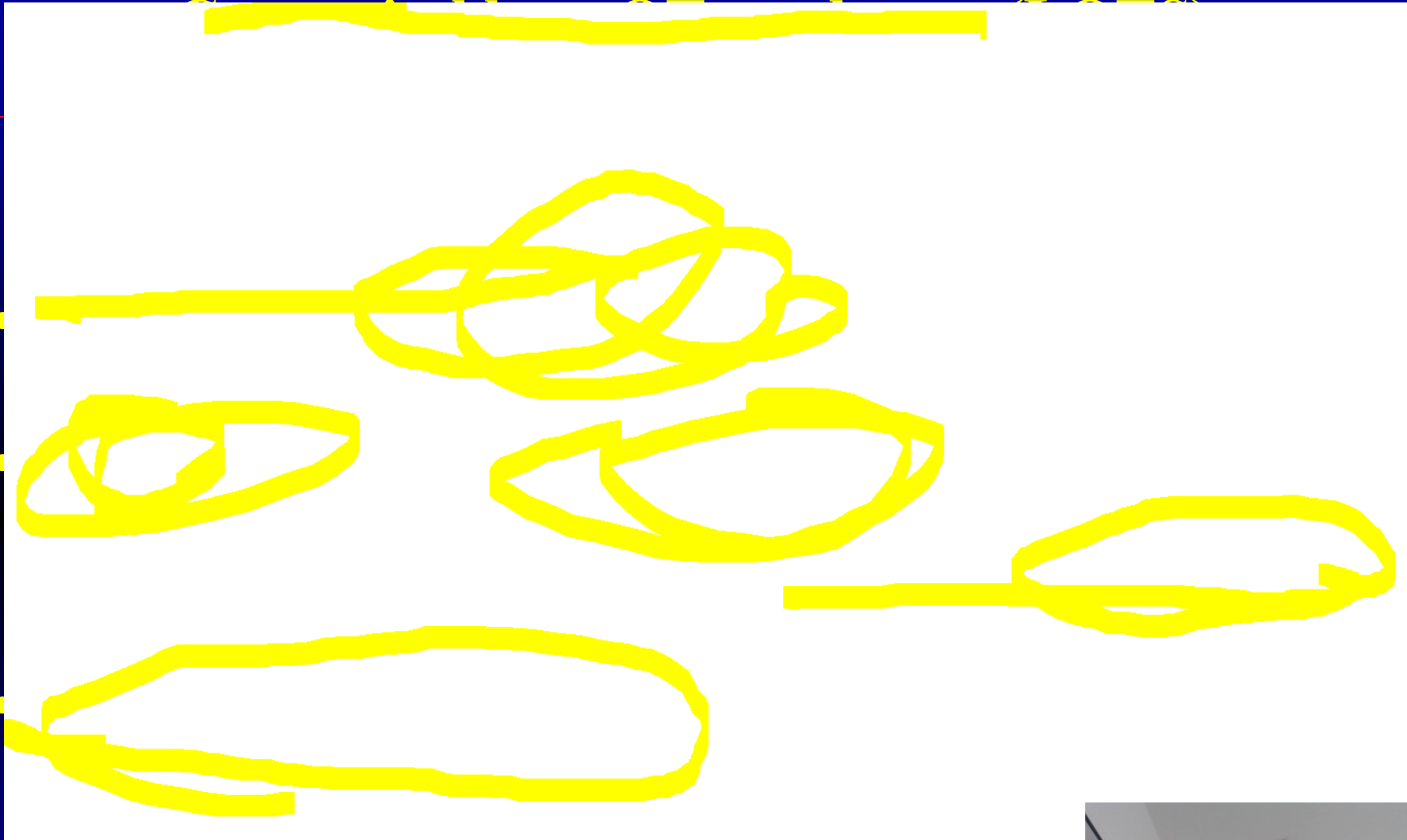
2) p[redominant features of atherogenic plaque] plaque
stab[le] coronary vessels

3) central and local control of myocardial excitability and vascular motorics



Hereditary arrhythmic diseases
=
a model of arrhythmogenesis







Ion channel genes

α subunit

β - subunit

SCN5A

SCN1B

KCND3, KCNA4

CACNA1A

CACNB1, B2

KCNH2

KC

KCNQ1

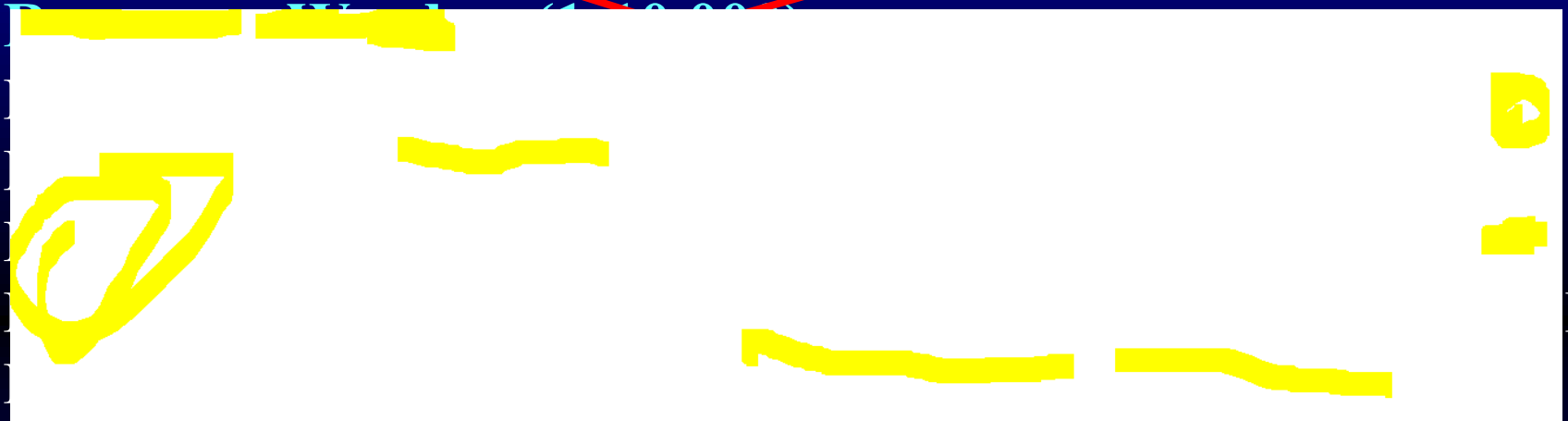
KC

KCNJ2, KCNJ12



LQTS types - genes and proteins

LQTS typ chromozom gen protein



LQT 6
(Andersen)

Jerven - 1

JLN 1

JLN 2

whites). In infants with a corrected QT interval (QTc) >450 ms, the ECG was repeated within 1 to 2 weeks. Genetic analysis, by screening 7 LQTS genes, was performed in 28 of 31 (90%) and in 14 of 28 infants (50%) with, respectively, a QTc >470 ms or between 461 and 470 ms. A QTc of 451 to 460, 461 to 470, and >470 ms was observed in 177 (0.41%), 28 (0.06%), and 31 infants (0.07%). Among genotyped infants, disease-causing mutations were found in 12

B
I_{Ks})
IRP (β I_{Kr})

prevalence of at least 1:2534 apparently healthy live births

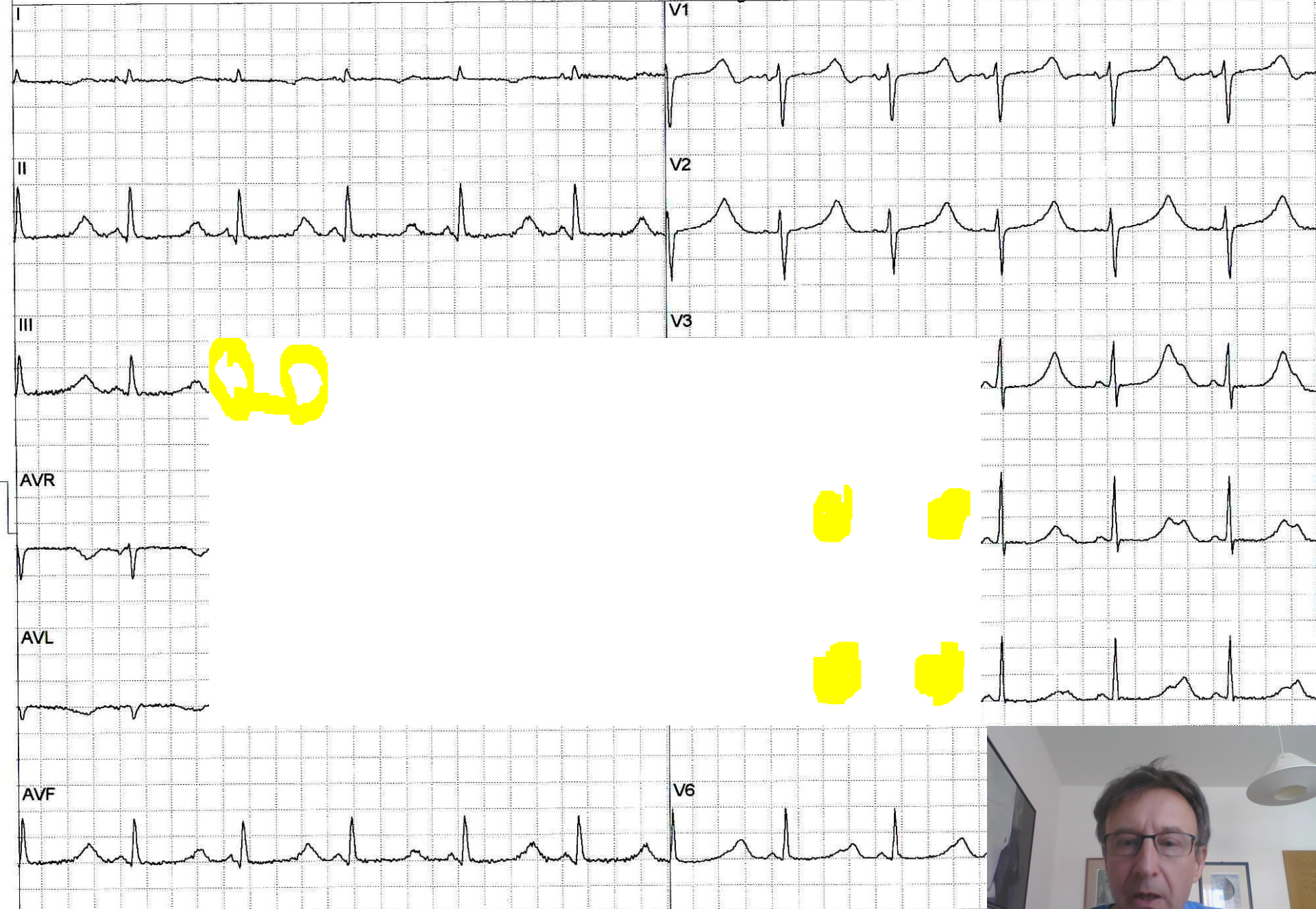
Conclusions—This study provides the first data-based estimate of the prevalence of LQTS among whites. On the basis of the nongenotyped infants with QTc betw close to 1:2000. ECG-guided molecular relatives, thus allowing effective preven *Circulation 2009;120:1761-*

21q22.1-22.2 *KCNE1*



LQTS diagnostic score (Schwartz et al., 1993)



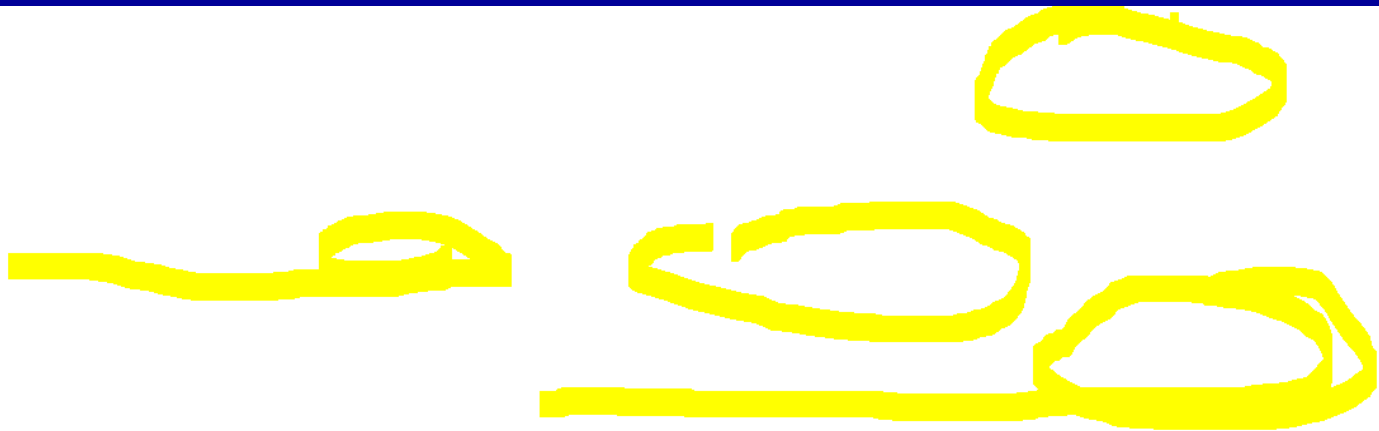




Batchvarov VN, et al. Am J Physiol Heart Circ Physiol 2000;282:H2







Rastr bez měřítka. HR 189 73 / 54 (65)

Torsade is an incomplete circulatory arrest
non-coordinated muscular activity – see



A case report



WCT

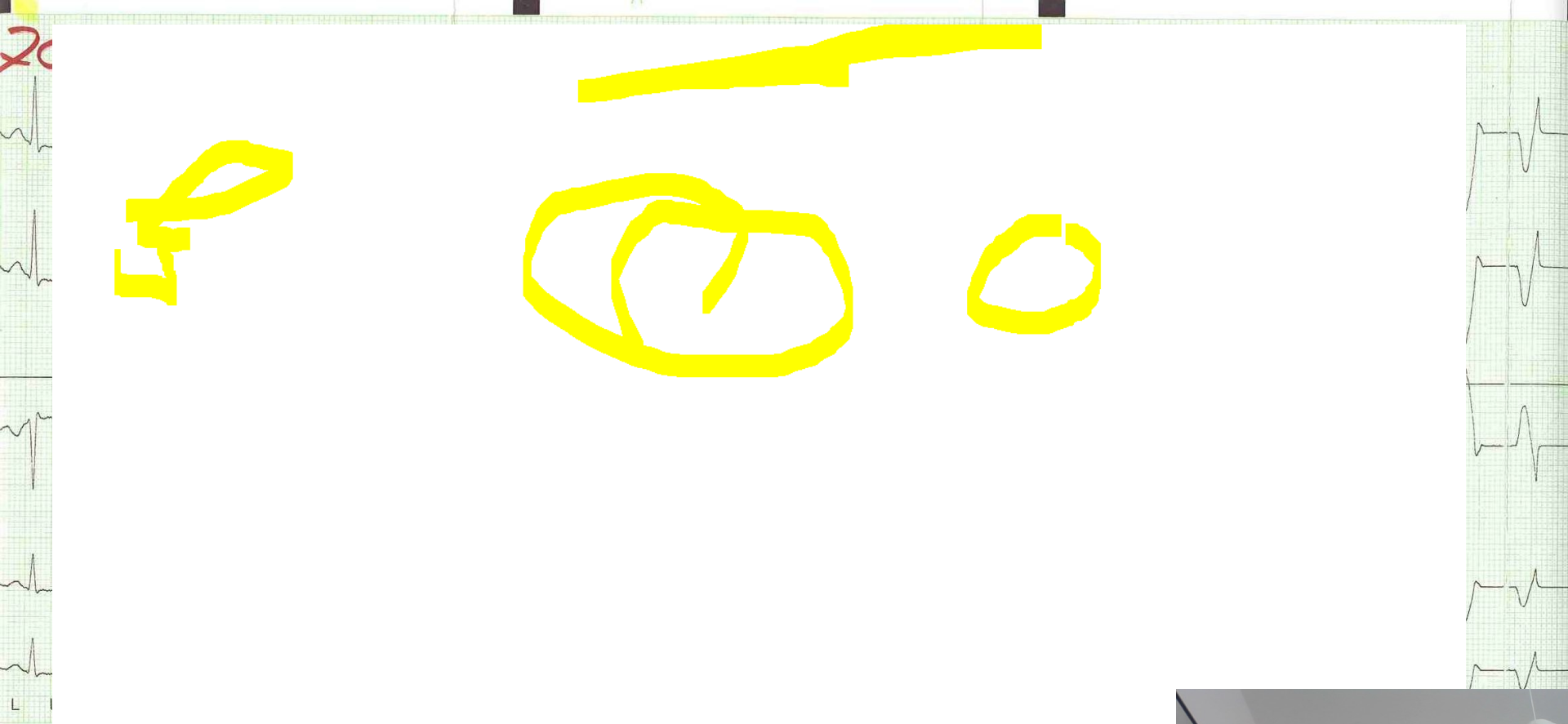
One normal

another normal

Mc



Continuing ECG shows QT interval prolongation



QT interval prolonging drugs

ajmaline, amiodarone, bretylium, dofetilide,
ide, ibutilide, procainamide, propafenone, chinidin,

therapeutics, antimycotics - amantadine,
ycine, chloroquine, cotrimoxazole, erythromycine,
e, halofantrine, itraconazole, ketoconazole, pentamidine,
iramycine, sparfloxacin

astemizole, loratadine, terfenadine

amitryptiline, clomipramine, clozapine,

chlorpromazine, citalopram, desipramine, doxepine, droperidol,

fl

m

p

thioniazine, umiperone, iminoperazone, veniarax

ziprasidone

www.qtdrugs.org

Other - cisapride, indapamide, ketanserine, probucol, hydro

sildenafil, vasopresin

