LONG QT SYNDROME AND ELECTRICAL STORM IN A SMALL BOY

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M.Sz

Born from the first pregnancy /in vitro/ of young parents by cesarean section at 32 hbd with a body weight of 1850 g.

Prenatally from the 23th week of gestation bradycardia with complete heart block was diagnosed, then tachycardia attacks were observed, the child was hemodynamically unstable, with increasing symptoms of heart failure, amiodaron was given to his mother with no effect.

During the first hours of his life the neonate was admitted to our cardiology department

ECG – bradycardia 60 bpm, complete heart block with wide QRS complexes.

Echocardiographic normal anatomy with left ventricular hipertrophy

M.Sz

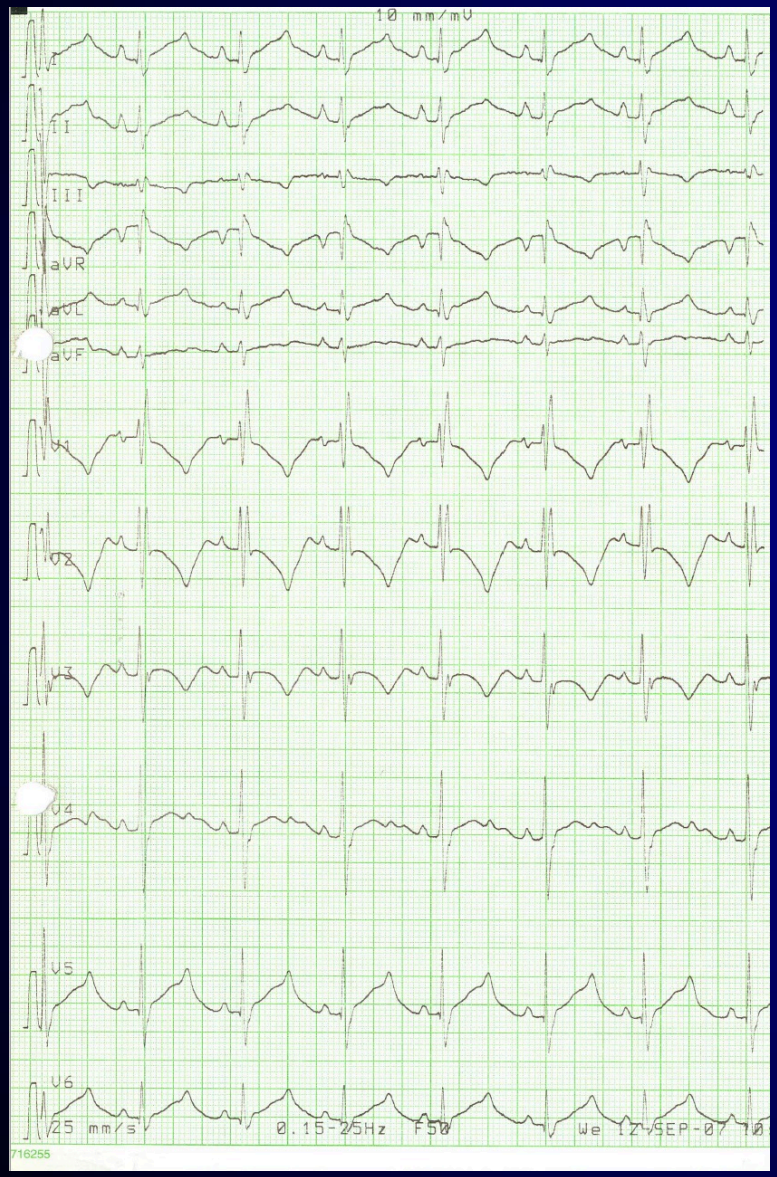
At his 2nd week of life, weight 1650 g, epicardial pacemaker system was implanted

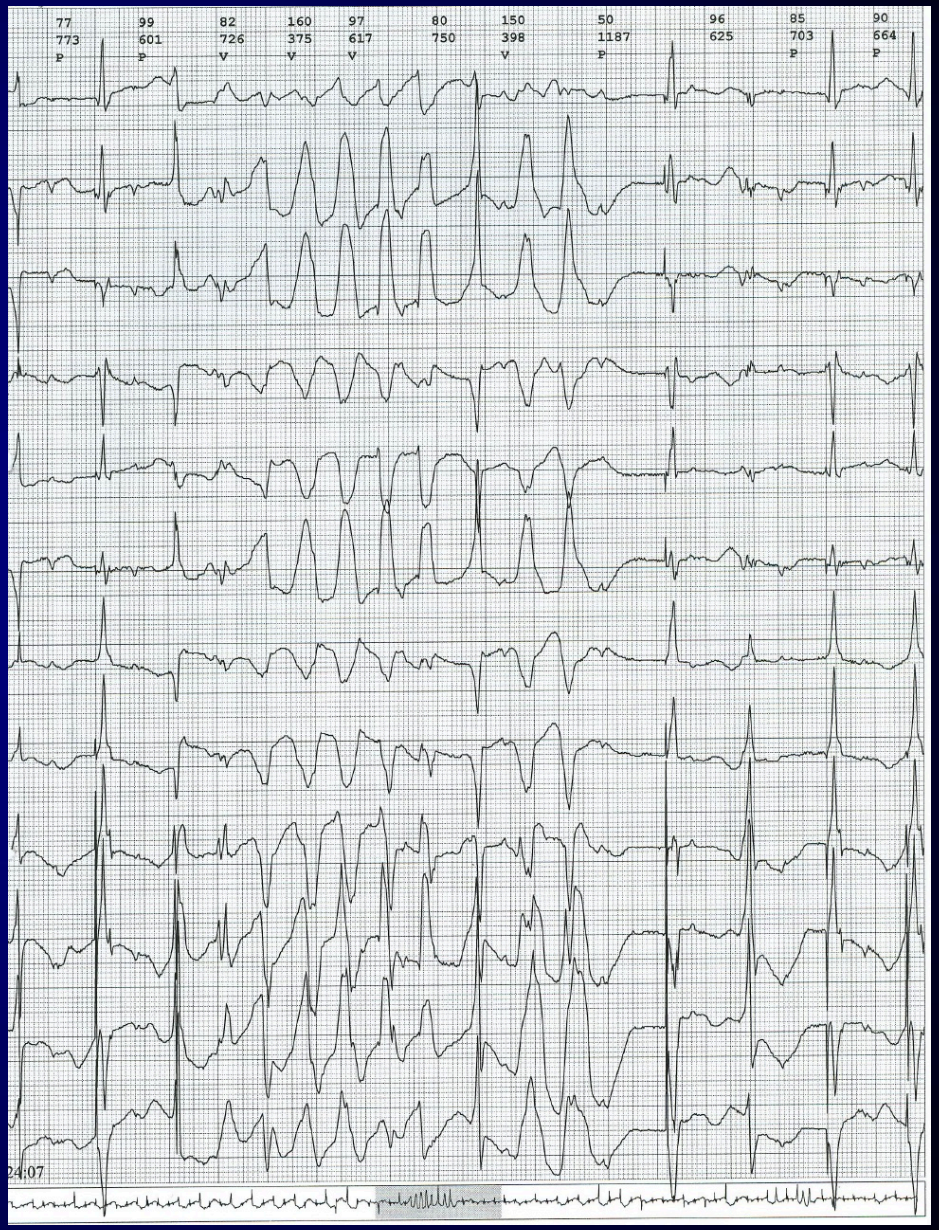
The child was discharged on low dose of propranolol due to prenatal tachycardia

Pacemaker function was correct

At age of 2 years 5 months he presented with syncope and was admitted to our department

On Holter ECG polymorphic ventricular arrhythmia was registered





There was QT prolongation on ECG

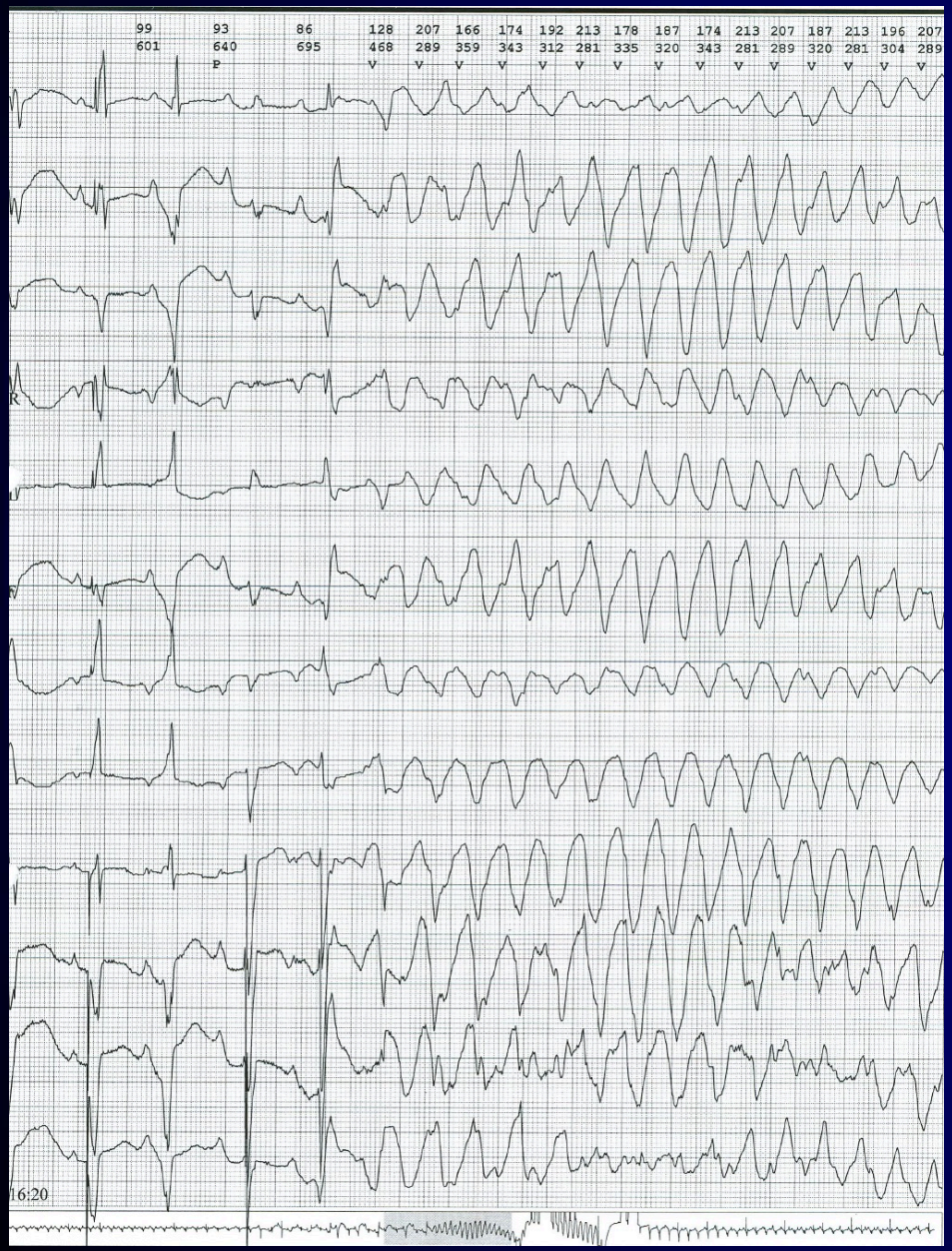
On Holter ECG polymorphic ventricular arrhythmia was registered

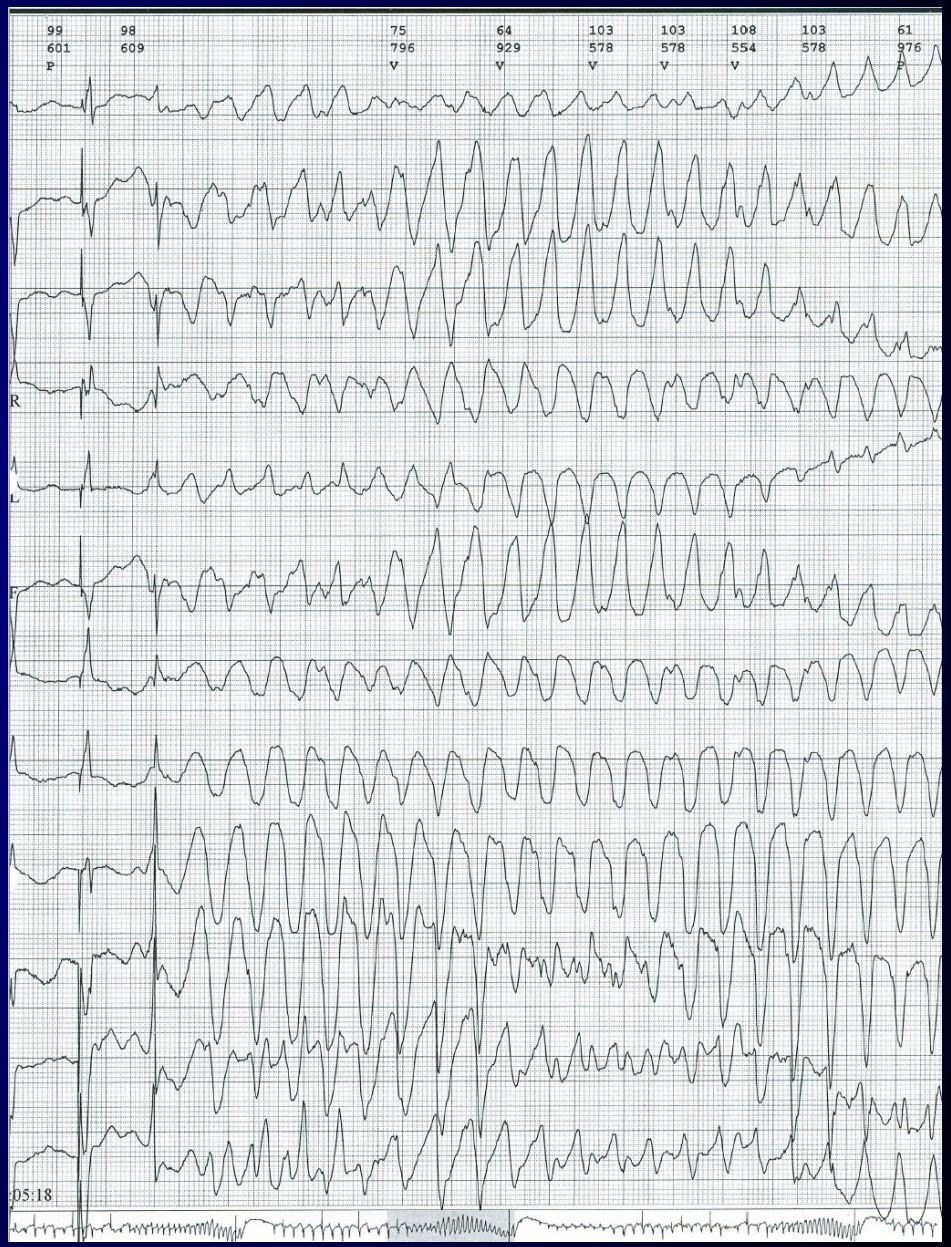
Pacemaker control revealed battery depletion

He was put on higher dose of Propranolol and pacemaker was changed

Immediately post surgery the polymorphic ventricular tachycardia episodes intensified

Life threatening polymorphic ventricular arrhythmia was registered





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We convereded pacemaker to epicardial ICD

ICD was programmed for VT and VF therapy

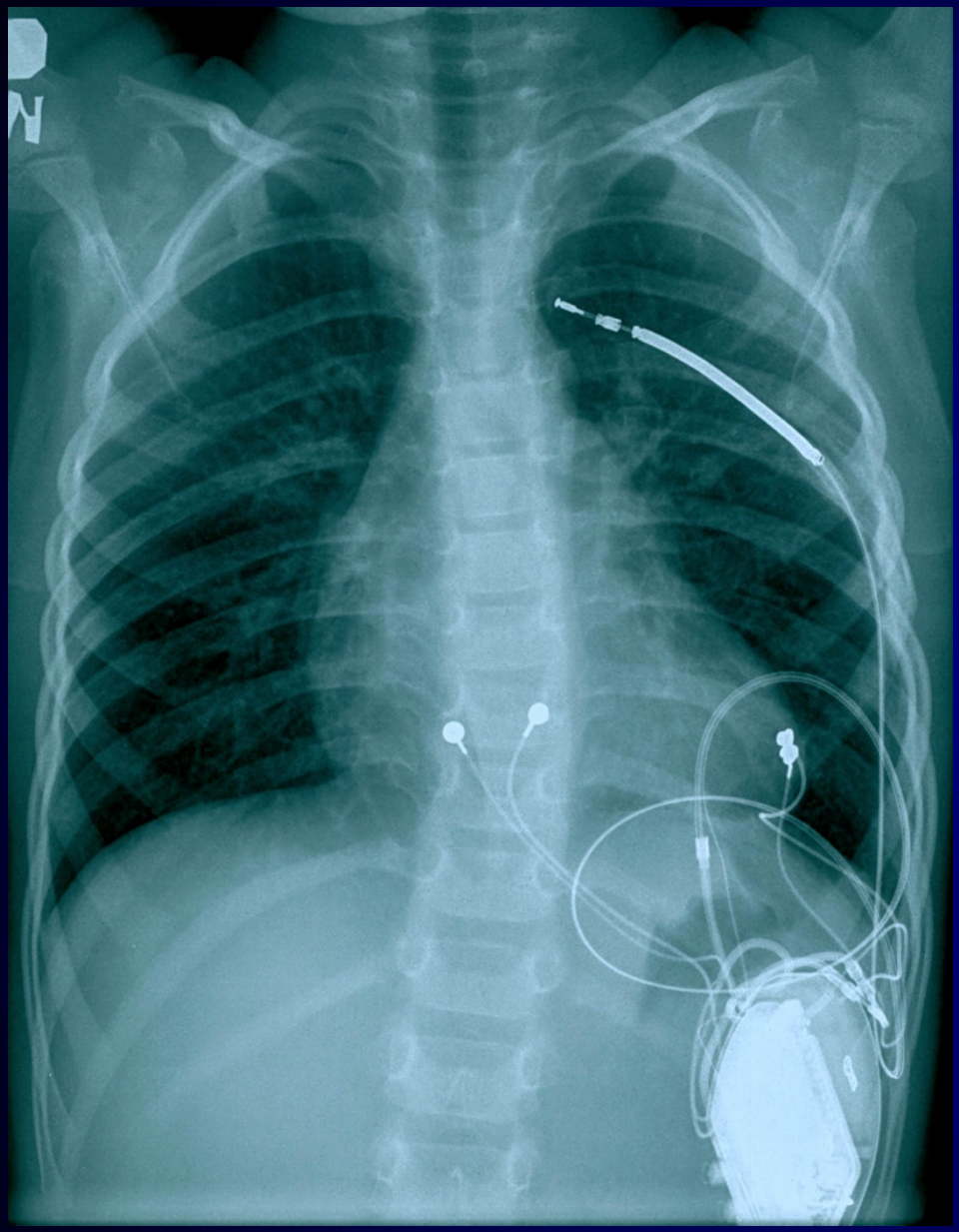
He received several shocks and started to complain from abdominal pain

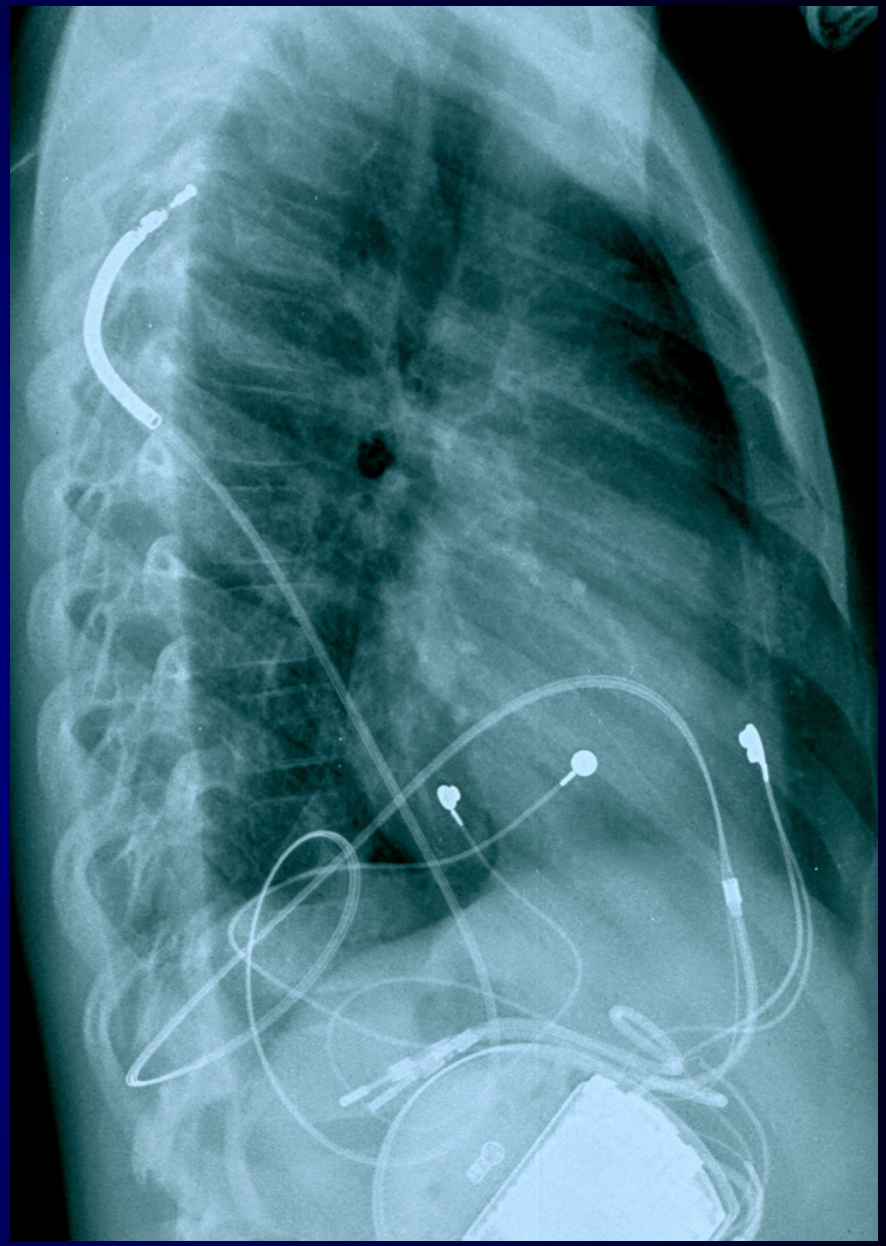
The VF episodes started to be longer and more frequent

The boy because of the electric storm was trasverred to ICU

He was sedated, intubated, mechanically ventilated and cooled to 35 C

Lidocaine and propranolol in very high doses were continouslly administered





2 years 6 months old,

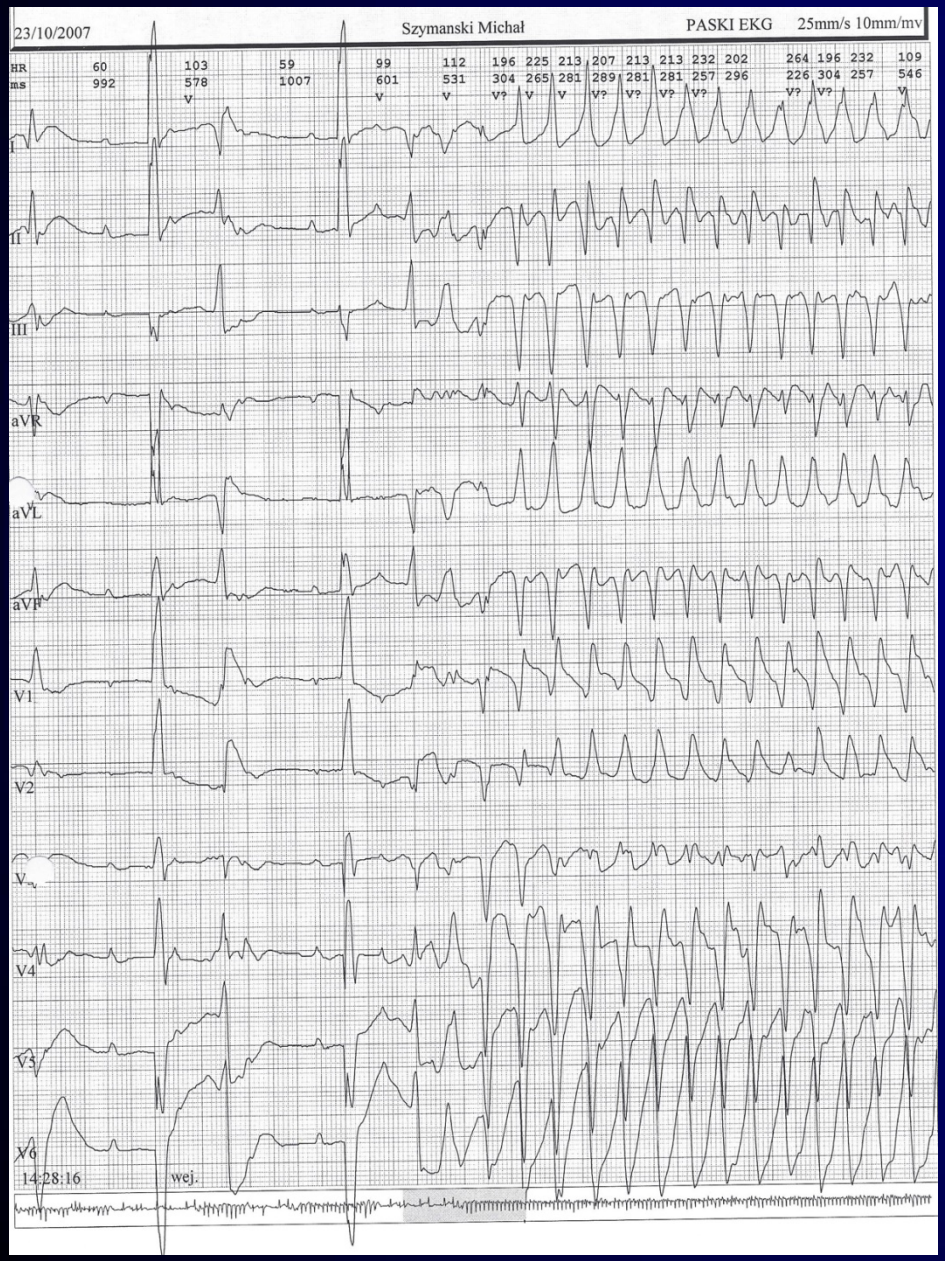
M.Sz

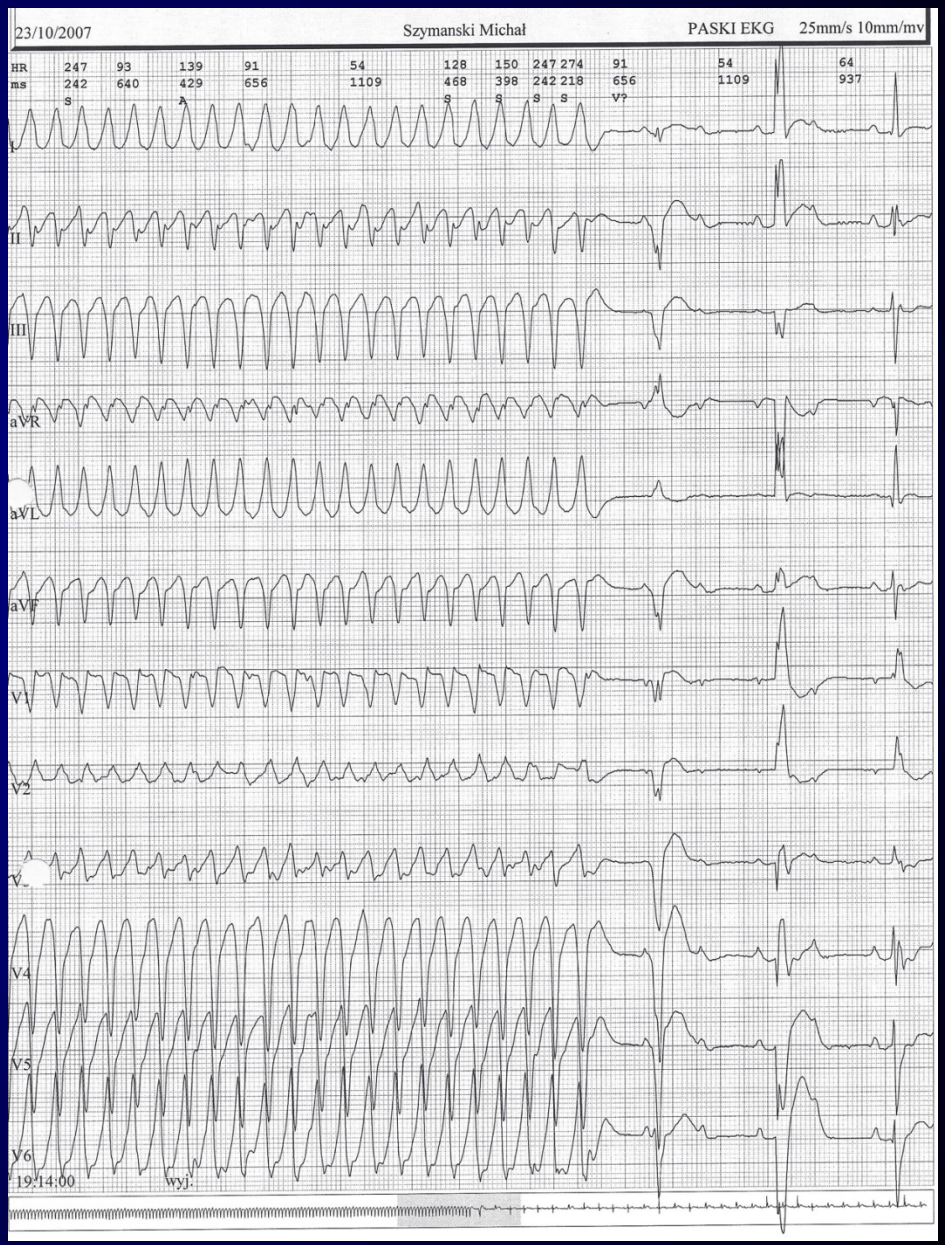
Life-threatening polymorphic VT still returned

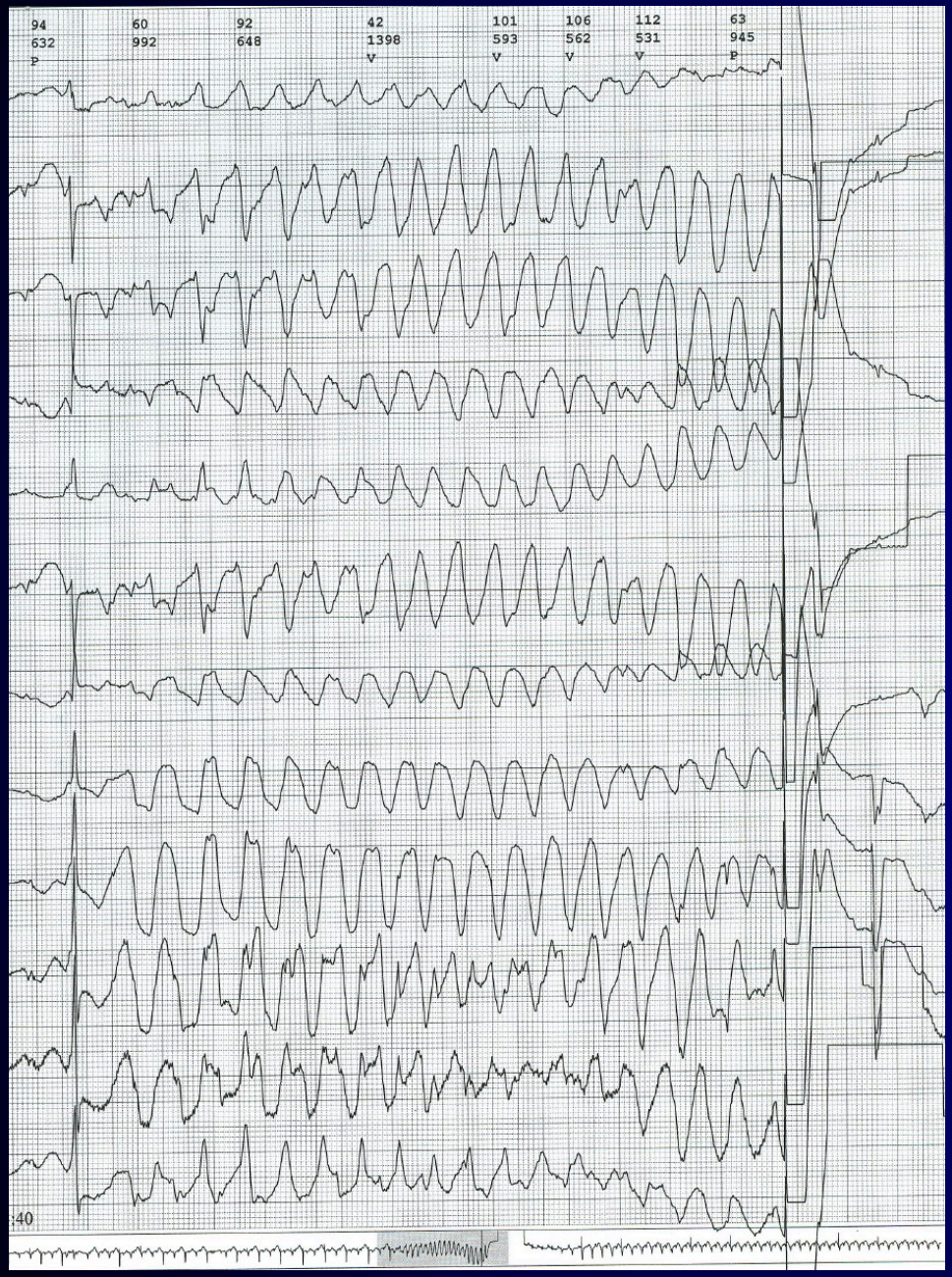
He had plenty of ICD therapies

We reprogramed ICD to stop therapies for some days.

M.Sz 3 weeks post ICD implantation, on propranolol and mexiletine



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63
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M.Sz.

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He improved very slowly, we rewarmed him after 5 days of body low temperature

We had to change ICD because of battery depletion after 25 days post its implantation

The patient was discharged on propranolol, mexiletine and aldacton

Long QT2 was diagnosed with HERG mutation