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Registry for cardio-cerebro-vascular pathology and sudden death in the young at the Veneto Region (North-East of Italy)

Registro de enfermedad cardio-cerebro-vascular y muerte súbita juvenil en la región de Véneto (Noreste de Italia)

Cristina Basso✉, MD; Stefania Rizzo, MD; and Gaetano Thiene, MD

Department of Cardiac, Thoracic and Vascular Sciences, University of Padua. Padua, Italy.

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To the Editor:

The story of the Veneto Region Registry, in Italy, on Juvenile sudden death (SD) dates back in the late 70s, when a series of SDs in the young occurred in the Veneto Region, North East of Italy. A successful application for a prospective study on young people dying suddenly (<35 years old, sudden infant death syndrome excluded) was submitted to the regional health authorities, thus implementing a network of collaboration with anatomic and forensic pathologists, to collect all such events and to gather epidemiological data. The Veneto Region is located in the North East of Italy with Venice as the capital. According to the Italian Census Bureau & Sports Medicine (1979-1999) the overall inhabitants was 4,379,00 – with the young population (12-35 years) of 1,386,650 and young athletes of 112,790 (M to F = 4:1). In this time interval the cumulative incidence of SD was 1/100,000/year in young people aged less than 35 years old (SIDS excluded). Among non-athletes, the incidence was 0.9/100,000/year and among athletes was 2.3/100,000/year (SD occurrence 2.5 fold in athletes vs non-athletes, p<0.0001).

In the time interval (1980-2013) a total of 650 SCD were studied at postmortem (201 F, 31%): it was mechanical in 7% and arrhythmic in 93%. Among the mechanical causes, pulmonary embolism accounted for 2%, aortic rupture for 3% and others in 1%. A congenital or genetic substrate was always there: Marfan syndrome, bicuspid aortic valve, aortic coarctation with or without bicuspid aortic valve. Coronary atherosclerosis was the cause of SCD in 18% of cases, usually with a single obstructive atherosclerotic plaque located in proximal descending coronary artery. Acute occlusive thrombosis was present in 34% in this group, mostly due to endothelial erosion. In 2% of cases, the acquired coronary artery disease was coronary dissection, usually in women. Congenital coronary artery abnormalities were the culprit in 5% of cases mostly due to anomalous origin from the wrong sinus. Myocarditis was diagnosed as

✉ *C Basso*
Department of Cardiac, Thoracic and Vascular Sciences, University of Padua.
Via 8 Febbraio 1848, 2, 35122 Padova PD, Italia.
E-mail address: revista.corsalud@gmail.com

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the cause of death in 12%. Arrhythmogenic cardiomyopathy resulted to be the third cause accounting for 10%. The fibro-fatty replacement was frequently biventricular and even isolated in the left ventricle or segmental in the right ventricle. Hypertrophic cardiomyopathy, with asymmetric subaortic, mid-ventricular or apical hypertrophy, accounted for 9%. Severe myocardial disarray and fibrotic scars, located within the asymmetric hypertrophy, were constant findings. Mitral valve prolapse was the only structural disease in 8% of subjects dying suddenly, largely prevalent in female gender. Fibrosis of the papillary muscles and of the postero-lateral free wall of the left ventricle was the most likely arrhythmogenic substrate. Conduction system disease, investigated by serial sections technique, was the cause of SD in 6%. In the majority, it was an ECG proven Wolff-Parkinson-White or Lown-Ganong-Levine ventricular pre-excitation. Congenital heart disease were observed in 2% of cases.

Finally, SCD occurred in young subjects in whom the hearts appeared normal at gross and histological examination (17%). Molecular autopsy was employed in the recent years and allowed to reveal pathogen genetic mutations in cases of long/short QT, Brugada and polymorphic ventricular tachycardia syndromes. As far as the athletes, arrhythmogenic cardiomyopathy accounted for 23% of SD victims, coronary atherosclerosis for 19%, congenital coronary artery anomalies for 16% and hypertrophic cardiomyopathy for only 2%. The discrepancies, when compared with the overall non-athletes population, have to be ascribed to the role played by effort in precipitating sudden death. Disqualification from sport activity is life-saving and this is the reason why in Italy rate of SD in athletes due to HCM is only 2% vs 26% in USA, a country where the pre-participation screening does not include the use of ECG. In the Veneto Region of Italy, in the time interval 1979 to 2004, the annual incidence of SD declined by 89% in screened athletes (P for trend <0.001).

In conclusion, SCD prevention in the young has to be faced by an interdisciplinary team, including pathologists, cardiologists, sport physicians, and geneticists, with a translational approach; the clinicopathologic correlation method still being the polar-star. In other words, the game in the fight against SCD is still played in the anatomical theater, the place where «death enjoys saving lives».

CONFLICT S OF INTEREST

None

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