Case Reports

Familial Polytopic and Polymorphic Extrasystoles

Erich KUHN, Dr. med., Priv. Doz., Dieter WOLF,
Dr. med., and Märgen STIELER, Dr. med.

IN 1960 an article from K. J. Berg appeared in *Am. Heart Journal*, entitled "Multifocal ventricular extrasystoles with Adams-Stokes syndrome in siblings." We have also observed the familial occurrence of extrasystoles and would like to offer as evidence the case histories of 2 sisters. A brother who succumbed to a sudden heart attack at the age of 10, and the mother, who died under similar circumstances at 40, very likely suffered from the same disturbance of rhythm. We must accept the word of their physician for this as the electrocardiographic findings were unfortunately lost during World War II. The father, one sister and three half-sisters (offspring of the father) are not affected,

Case 1. Gisela D., born July 14, 1943, the younger sister, was first observed to have abnormal cardiac findings at the age of 11 years. She was admitted to the University Pediatric Clinic at 14 with angina pectoris. There was no history of rheumatic fever or diphtheria. Two years later she was examined by us because of frequent syncopal attacks.

On physical examination at the age of 15 she was pale and poorly developed, 5 ft, 4 ins tall and weighed 105 pounds. She had a small soft goitre and the pulsations of her carotid vessels were more obvious than normal. A grade III systolic murmur was noted over the entire precordium.

The phonocardiogram showed the murmur to be protosystolic and of moderate frequency. Attacks of tachycardia with frequency up to 200/min. occurred. No other significant abnormalities were found. Laboratory studies included a normal blood picture, urinalysis and a serum potassium of 5.1 mEq./L.

The chest roentgenogram was normal. The roentgenogram of the skull showed absence of the upper lateral incisors and the third molars. In and the electrocardiogram, one or more ventricular extrasystoles follow each normal beat. Sometimes A-V premature beats appear in the postextrasystolic pauses. Sometimes there is a bigemini or trigemini form. Short runs of 3, 4 or 5 extrasystoles are quite frequent even at rest (Fig. 1a). In the normal beats there is a flattening of the T in V_6 (Fig. 1b and c).

From Medizinische Universitäts-Poliklinik Heidelberg (Direktor: Prof. Dr. H. Plügge)
and Universitäts-Kinderklinik Heidelberg (Direktor: Prof. Dr. Ph. Bamberger)

Fig. 1 a to d. Electrocardiograms of Gisela D. during 1957 recorded. An interpretation is included in the text. Tracings at later dates show the same picture.

Often the form of the individual extrasystoles and their temporal relationship repeat after hours or days. The frequency of the extrasystoles tends to increase with exertion or even with standing. Then there appear attacks of tachycardia of varying duration with a frequency of 180/min. (Fig. 1d). The attacks usually end after some seconds but also not infrequently may continue for minutes, hours, or even days and then lead to syncopal attacks. QRS complexes show alteration in form with constant ventricular interval. P waves are not to be seen with any certainty. An impulse originating focus in the left ventricle is probably responsible for the origin of the extrasystoles. Holzmann⁴⁾ explains this form of ventricular tachycardia by means of an altering refractory condition of a myocardial area which borders on the origin of the impulse or even lies more distant from it.

Case 2. Gerda D., born April 23, 1937, who is about 6 years older, is obviously similar in facial features and body build to her younger sister, and like her is missing the upper lateral incisors. She complains she is short of breath, has chest pain, and tires easily. She suffers occasionally from tonsillitis but has never been seriously ill. The physical findings, including heart size, are normal. The blood pressure is 125/75. She has tachycardia of 100/min. and extrasystoles.

The electrocardiogram shows a marked non-phasic sinus arrhythmia. Numerous polytopic and polymorphic ventricular extrasystoles are to be seen. These are either irregularly distributed within the fundamental rhythm or also

Fig. 2 a and b. Electrocardiograms of Gerda D. recorded during 1959. An interpretation is included in the text. Tracings at later dates show the same picture.

occur in bigeminal or trigeminal form (Fig. 2a). The extrasystoles chiefly arise in the left ventricle. The postextrasystolic pauses are fully compensated. On exertion the sinus frequency increases to 150/min. (Fig. 2b). To be noted are the broad peaked P waves in Lead II, the depression of ST in II and III and the flattening of T in III. Only a few seconds after beginning exertion ventricular extrasystoles appear. The coupling intervals are shorter than at rest. A bigeminy appears first, which is interrupted occasionally by a trigeminy. Several minutes after exertion the frequency of the extrasystoles decreases again. In 1960 after a normal pregnancy she bore a full term male infant. The child has club feet but otherwise the child is normal.

DISCUSSION

The cases of 2 sisters in which polymorphic and polytopic ventricular extrasystoles occur have been presented. The extrasystoles show a fixed coupling for a time and then fall into bigeminy or trigeminy. On exertion the coupling intervals decrease. At rest the frequency of the extrasystoles may increase without known cause; in standing and on exertion it always increases. In the case of Gisela D. may lead to paroxysmal tachycardia with syncopal attacks and may be considered to be "tachycardie paroxystique à excitable" as was described by Gallavardin.²⁾

The mother of the 2 sisters died of sudden heart failure at the age of 40 years, and a brother of the same cause at 10 years. A third sister is said to be healthy. She has not yet however been thoroughly examined. The son of Gerda D., who is now about one year old, does not yet have extrasystoles and his electrocardiogram is normal. We do not however know whether the extrasystoles in his mother and aunt were present at birth or developed later, as we ourselves have observed the development of left bundle branch block in a patient with myotonia dystrophica.⁵⁾ Thus it can not yet be said whether he suffers from the gene defect—for such a defect most likely exists in his mother and aunt—which only becomes manifest at later stage of development. In myotonia dystrophica in which disturbances of heart rhythm including extrasystoles may occur,^{1), 3), 6)} a genetic tendency in this direction may only later become manifest. What sort of a gene is present in our patients must at the present time be left entirely open.

SUMMARY

The cases of 2 sisters with polymorphic and polytopic ventricular extrasystoles are reported. One has syncopal attacks particularly in combination with "tachycardie paroxystique à excitable" as was described by Gallavardin. The mother and a brother succumbed to sudden heart

attack very likely as a result of the same disturbance.

ACKNOWLEDGMENT

We are greatly indebted to Dr. John Wild for a critical review of the manuscript and to Dr. Ellen Monkus for assistance with the translation.

REFERENCES

1. Fisch, Ch.: Am. Heart J. **41**: 525, 1951.
2. Gallavardin, L.: Arch. mal. cour, Paris, **15**: 167, 1922.
3. Griffith, T.W.: Quart. J. Med. **5**: 229, 1911.
4. Holzmann, M.: Handbuch der Inneren Medizin, Berlin-Göttingen-Heidelberg, IX/2, 28 1960.
5. Kuhn, E.: Schweiz. Med. Wschr. **90**: 1160, 1960.
6. Kuhn, E.: Ztschr. f. Kreislauffschg. **50**: 149, 1961.