First- and Second-Degree Atrioventricular Block

15

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The primary role of the AVN is to conduct electrical action potentials from the atria to the ventricles. The AV node introduces a delay in the conduction of the electrical impulse from the atria to the ventricles to allow atrial mechanical systole to complete the filling of the ventricles before ventricular systole. The His bundle, in continuity with the AV node, provides an escape (backup) pacemaker (albeit much slower) in case of sinus node failure.

Anatomy and Electrophysiology of the AV Node

Electrical conduction through the myocardium is dependent on the individual myocyte, cell-to-cell conduction, and propagation through the whole organ. One cellular characteristic that contributes to impulse conduction velocity in the heart is the composition and density of specific ion channels that are responsible for cellular transmembrane depolarization. The more rapidly a local region of the cell membrane is able to change its potential (inside relative to the outside), the more rapid is conduction down the length of the myocyte. Therefore, cells that depend on the rapid sodium current (such as the atria, ventricular, and His–Purkinje tissue) for the upstroke of phase 0 of the action potential conduct the signals rapidly, while cells that are dependent on the slower calcium current for phase 0 conduct signals more slowly. Although the various phases of the action potential of cell membranes are generated by more than one ion current, this difference is demonstrated by comparing the slower AV node conduction velocity (dominant calcium-dependent action potentials) to the faster atrial and ventricular tissue conduction velocity (dominant sodium channel-dependent action potentials). Another property that contributes to conduction velocity is the nature of the cell-to-cell connections—the gap junctions composed of different expressions of connexin membrane proteins. Connexin proteins cluster at gap junctions and form intracellular channels between cardiac myocytes. Electrical coupling in the AV node is poor because there are few gap junctions and the gap junctions are small. There are four connexin protein isoforms expressed within the heart (C×40, C×43, C×45, and C×30.2); their distribution in the different tissues within the atria, ventricles, and AV node vary and lead to the different electrical properties in each. Conduction through the AV node is slower than through atrial

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or ventricular myocardium for multiple reasons: lower concentration of Na+ channels in the AV node, contributing to the delay in conduction, a more complex arrangement in that they are separated by connective tissue and are also smaller in diameter, both contributing to slower conduction and the distribution of the different connexins in and around the compact AV node. In addition, the conduction through the AV node is strongly modulated by autonomic innervation to the node, a number of forms of cardiac abnormalities, as well as the patient's basal state.

The AV node is located in the atrial septum at the apex of the triangle of Koch and is connected to the His bundle; the AV node can be divided into the lower node bundle and the compact node. The AVN region comprises functionally separate fast and slow pathways. Recent studies examining the histologic and molecular structure (ion channels and connexin isoforms) of the AV node and His bundle indicate that there are different functional components of the AV node and the His bundle and that they have different conduction properties. A rightward (inferior node) extension likely corresponds to a slow (conducting) pathway; a leftward extension and the corresponding fast (conducting) pathway are less well established (see Chaps. 1 and 5).

Abnormal conduction to the ventricles can result from intrinsic AV nodal or infranodal (the His–Purkinje system) disease (all potentially, deleteriously altering the cellular structural and electrical properties of the AV conduction system) including inflammation, infection, and degenerative changes including cardiomyopathy and apoptosis, or from extrinsic causes, including abnormal autonomic tone, electrolyte imbalance, hypothermia, trauma (surgery), and medication effects. First-degree AV block is almost always due to abnormal conduction in the atrium or AV node. Up to 20 % of patients with first-degree AV block and congenital heart disease, most notably AV septal defect and Ebstein's anomaly, have, in addition, prolonged intra-atrial conduction. Transient prolongation or failure of AV conduction may result from concealed conduction of atrial, junctional, or ventricular extrasystoles (Figs. 15.1, 15.2, and 15.3). Second-degree AV block may occur within the AV node, the His–Purkinje system, or at the ventricular level.

Similar to the sinus node, the AV node is innervated by the autonomic nervous system, which consists of a complex interaction of the sympathetic and parasympathetic nervous systems.

Cardiac chronotropic and inotropy are influenced by differing sympathetic and parasympathetic effects. Sympathetic nerves descend from the brain to the stellate and paravertebral ganglia where they synapse with postganglionic neurons. The sympathetic nervous system increases heart rate and contractility by binding norepinephrine to adrenergic receptors, initiating the adenylate cyclase-cAMP cascade. Parasympathetic nerves originate in the brain stem and project from the vagus nerve to postganglionic fibers within cardiac ganglia. The parasympathetic nervous system slows heart rate and decreases contractility by binding acetylcholine to cardiac muscarinic receptors or neural nicotinic receptors. Postganglionic sympathetic nerves extend from

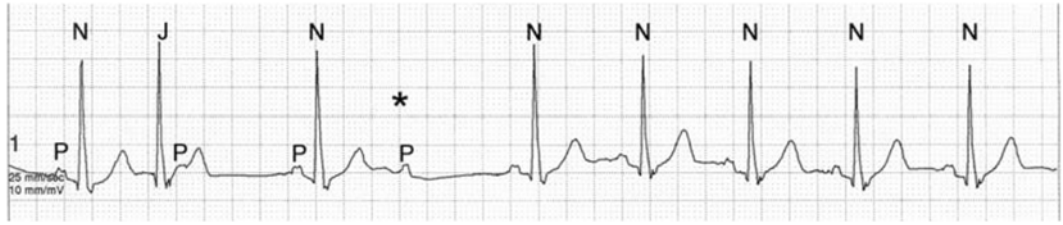

Fig. 15.1 Sinus rhythm with a junctional premature beat (J), which results in AV node refractoriness and block of the next sinus beat. *Subsequent non-conducted atrial impulse may be due to concealed junctional extrasystole, again making the AV node refractory without activating atrium or ventricle. Proof of this phenomenon requires intracardiac recordings

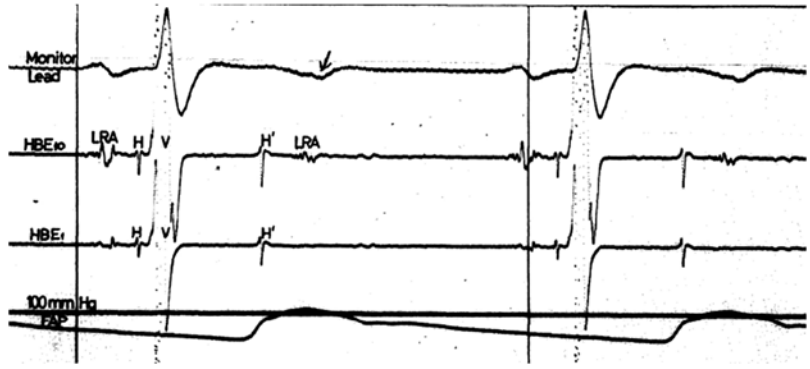

Fig. 15.2 Spontaneous His bundle extrasystoles (H′) arising at an H–H′ interval of 257 ms producing bigeminal rhythm. H′ fails to conduct to the ventricle but conducts retrograde to the atrium resulting in reversal of atrial activation from low to high atrium, associated with an inverted P wave (*arrow*). The bigeminal His bundle extrasystoles cause a marked slowing of ventricular rate. [Reprinted from Nasrallah AT, Gillette PC, Mullins CE, et al. Concealed his bundle extrasystoles in congenital heart disease. *American Journal of Cardiology*. 1975;35(2): 288–292. With permission from Elsevier.]

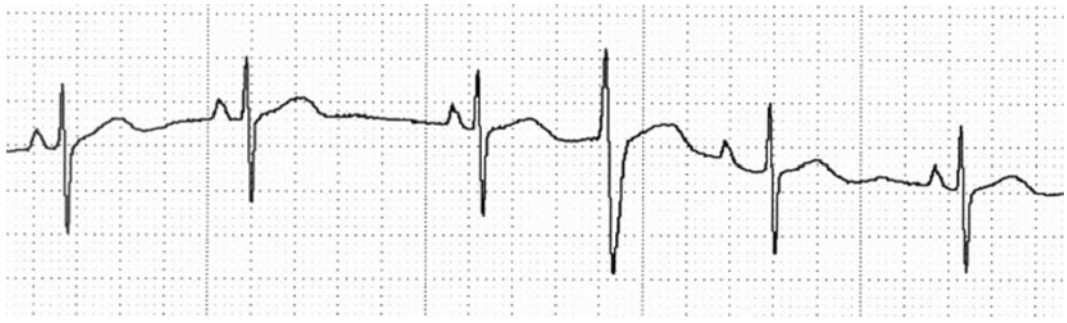

Fig. 15.3 Sinus rhythm with interpolated PVC. Concealed retrograde conduction into the AV node results in relative refractoriness and first-degree block (longer PR interval) of subsequent sinus impulse

the paravertebral ganglia to the heart where they meet with the parasympathetic nerves to form the cardiac neuronal plexus at the base of the heart.

The sympathetic innervation of the conduction system dominates in infancy but shifts to a balance of sympathetic and parasympathetic innervation in childhood and is complete in adulthood. Autonomic innervation is less prominent in the His–Purkinje system and has less influence on conduction in these areas. Bradycardia and PR interval prolongation usually reflects increased vagal tone on the sinus and AV nodes. PR interval prolongation in the presence of normal sinus rates is suggestive of AV nodal dysfunction. Functional first- and second-degree AV block can also present during rapid atrial pacing (Fig. 15.4), where, unlike sinus tachycardia, there is less sympathetic enhancement of AV nodal conduction.

ECG Characteristics

Multiple channel ECG recordings are often indispensable for detection of P wave morphology and PR intervals. Recording of atrial activity from esophageal or temporary epicardial pacing leads after surgery (Fig. 15.5) are also very useful when P waves are indistinct. Long-term recordings from Holter monitoring often reveal patterns not apparent in the 12 lead electrocardiogram.

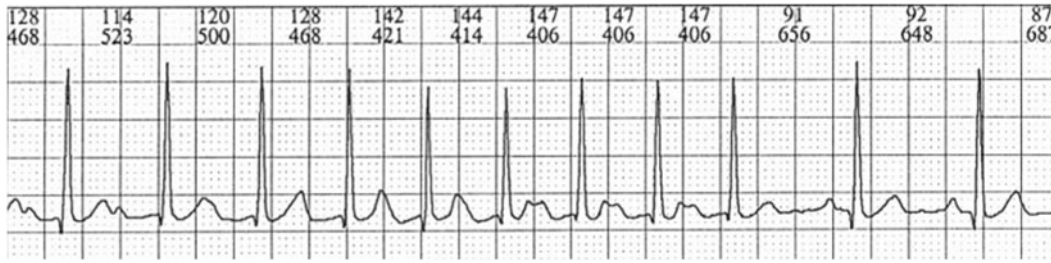

Fig. 15.4 Atrial pacing (stimulus artifact not present) producing a tachycardia that transitions to sinus rhythm. There is first-degree AV block during the tachycardia that resolves with the return of sinus rhythm

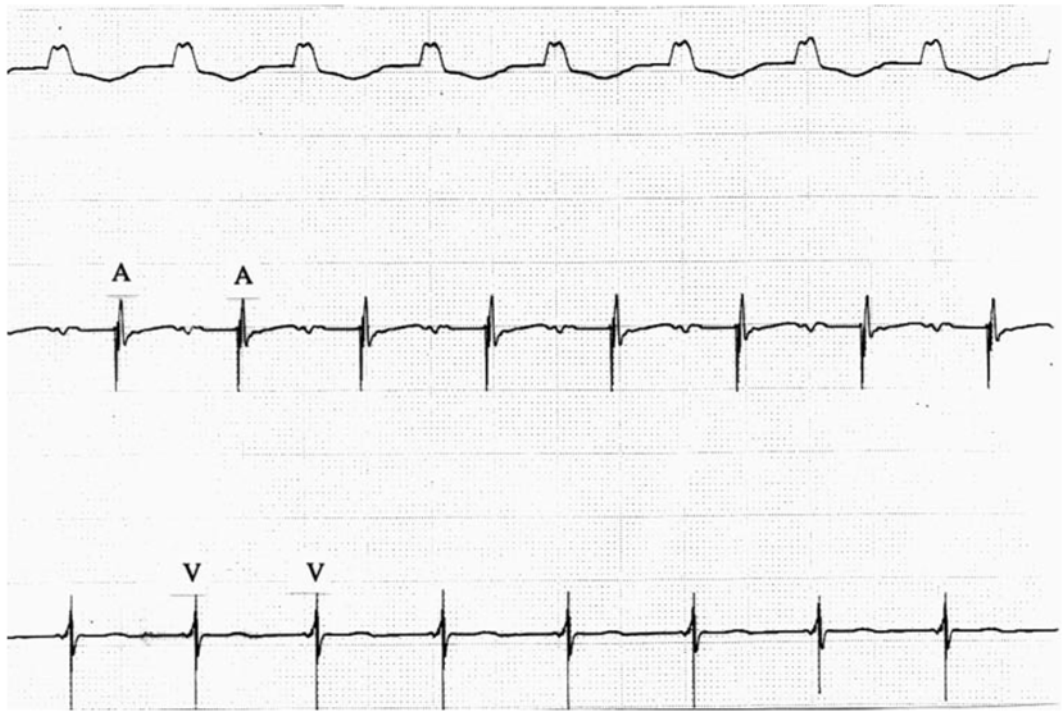

Fig. 15.5 Surface ECG (*upper signal*), atrial wire recording (*middle signal*), and ventricular wire recording in a patient with first-degree AV block following surgical repair of an atrioventricular septal defect. Atrial activity was not apparent from the surface ECG. Atrial wire recordings confirm 1:1 AV relationship with prolonged AV conduction time

First-Degree AV Block

First-degree AV block (Fig. 15.6) is defined as a PR interval above the normal range for age, but with persistent 1:1 AV conduction. The normal PR interval also decreases with increasing heart rate. Age-appropriate PR intervals are summarized in Table 15.1. The PR interval can be very long (Fig. 15.7) but usually, in the absence of heart disease, does not progress. The delay is located in the AV node mediated through excessive parasympathetic tone. Exercise, both recreational and during stress testing, induces parasympathetic withdrawal and sympathetic enhancement resulting in normalization of AV conduction and the PR interval (Fig. 15.7).

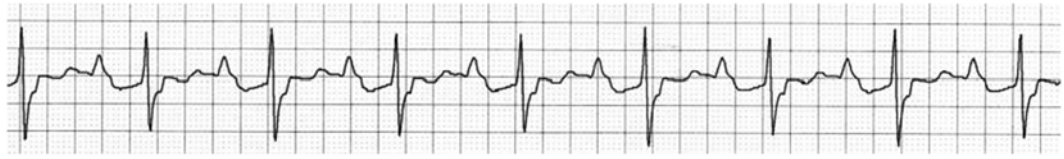

Fig. 15.6 Sinus rhythm with first-degree AV block

Table 15.1 Maximum normal PR interval

Age	PR (s)
0–3 days	0.16
4–30 days	0.14
1–3 months	0.13
4–6 months	0.15
7–12 months	0.16
1–5 years	0.16
6–12 years	0.17
>12 years	0.20

Second-Degree AV Block

Several distinct patterns of second-degree AV block can be recognized. Consistent periodicity (i.e., dropping every third, fourth, or fifth beat) is frequently present. The ratio of P-to-R waves provides a description of the pattern (i.e., 3:2, 4:3). This pattern of "grouped beats" should always suggest second-degree AV block. Regular non-conducted atrial extrasystoles may also present with this pattern and are distinguished by the irregular PP interval, as well as the different P wave morphologies (Fig. 15.8).

Mobitz I (Wenckebach)

With typical Mobitz I, or Wenckebach (Figs. 15.9 and 15.10), there is gradual prolongation of the PR interval prior to a non-conducted beat. The greatest increase in PR interval is between the first and second conducted beats of a series. The lesser increment increase of the PR interval on subsequent beats leads to a shortening of the RR interval. Following the non-conducted beat, the normal PR interval is restored resulting in an RR interval that is less than twice the sinus rate.

Atypical Wenckebach, which may be more common than the typical form, refers to other patterns of PR prolongation in association with the appearance of a dropped beat (no conduction to the ventricles)—AV block. This pattern may occur in the presence of sinus arrhythmia and with longer runs of conducted beats between block cycles.

Mobitz II

The PR interval does not vary prior to non-conducted beats with Mobitz II second-degree AV block, and, in the absence of a supraventricular arrhythmia, the RR interval is constant (Fig. 15.11). Since there is no change in PR or RR interval during the conducted beats, the RR interval following a non-conducted beat should be twice that of a conducted RR interval.

When every other beat is non-conducted in a 2:1 pattern, insufficient information is present to distinguish the pattern of Mobitz I from Mobitz II. Long-term recordings may reveal other ratios of conduction allowing discrimination of these two entities (Figs. 15.10 and 15.11). Long QT syndrome may present with 2:1 conduction as the ventricles, due to the mutated K+ ion channel (LQTS1) delaying ventricular repolarization, are refractory to successive sinus impulses; it is a poor prognostic sign.

Advanced

Advanced AV block is present when two or more impulses are not conducted in the absence of complete heart block (Fig. 15.12). Patients with advanced AV block may progress to complete heart block (Chap. 16).

Electrophysiologic Features

Electrophysiologic studies have provided invaluable insights into AV node physiology. Currently, clinical history and noninvasive diagnostic studies

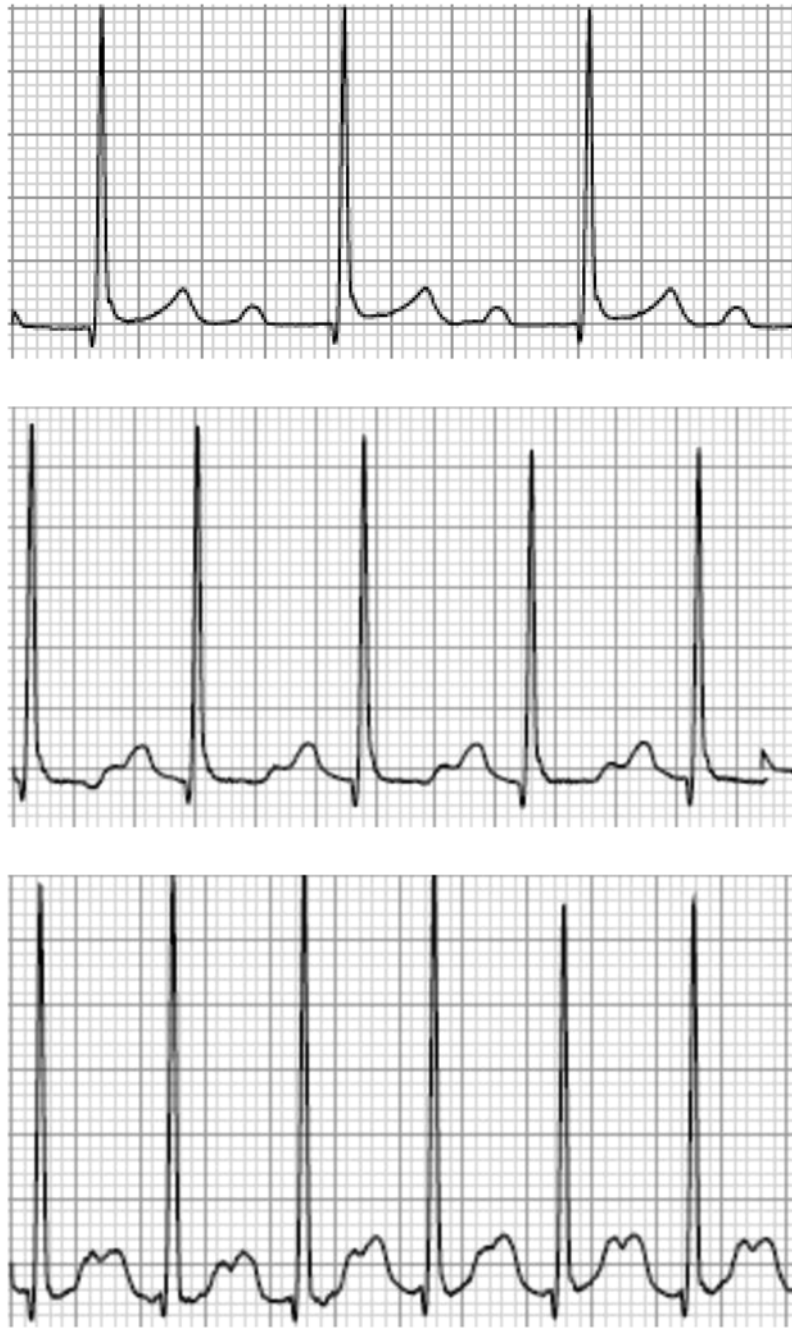

Fig. 15.7 16-year-old girl with first-degree AV block at baseline. With exercise, the PR interval shortens and there is 1:1 AV conduction

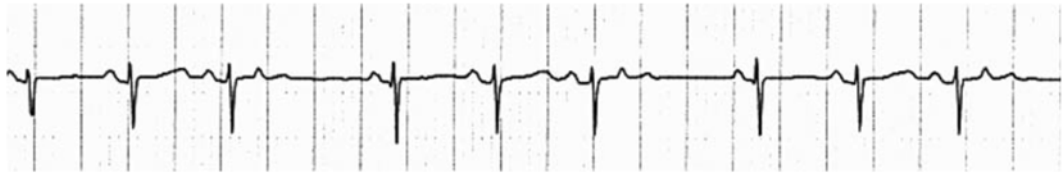

Fig. 15.8 Sinus rhythm with frequent non-conducted atrial extrasystoles in a quadrigeminal pattern. The grouped beats mimic second-degree AV block, but the variation in P wave timing and morphology are distinguishing features

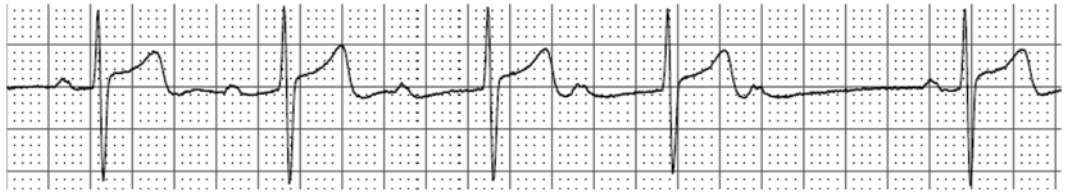

Fig. 15.9 Sinus rhythm with first-degree and Mobitz I second-degree AV block with a 5:4 conduction ratio

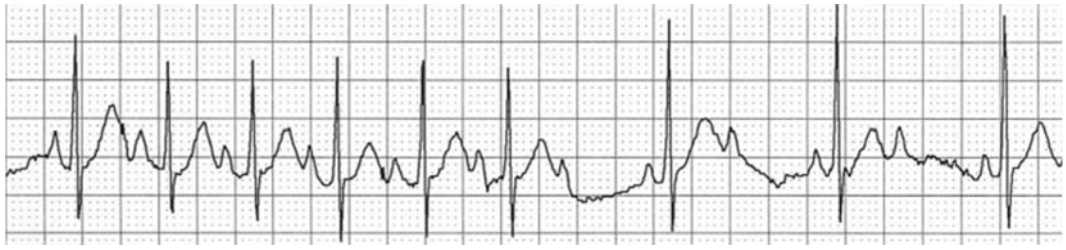

Fig. 15.10 Sinus rhythm with Mobitz I second-degree AV block that transitions to 2:1 block

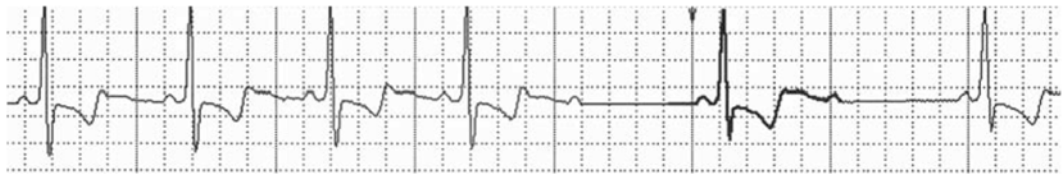

Fig. 15.11 Sinus rhythm with stable PR interval that transitions to 2:1 block. In the absence of prior PR interval prolongation, this most likely represents Mobitz II AV block

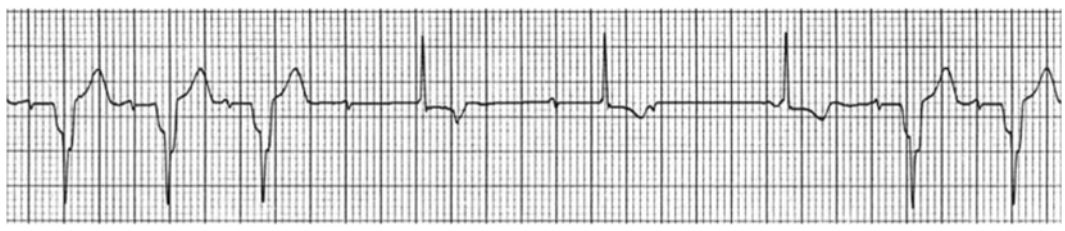

Fig. 15.12 Sinus rhythm in a patient with nodoventricular pathway. Advanced AV block associated with narrow complex escape rhythm

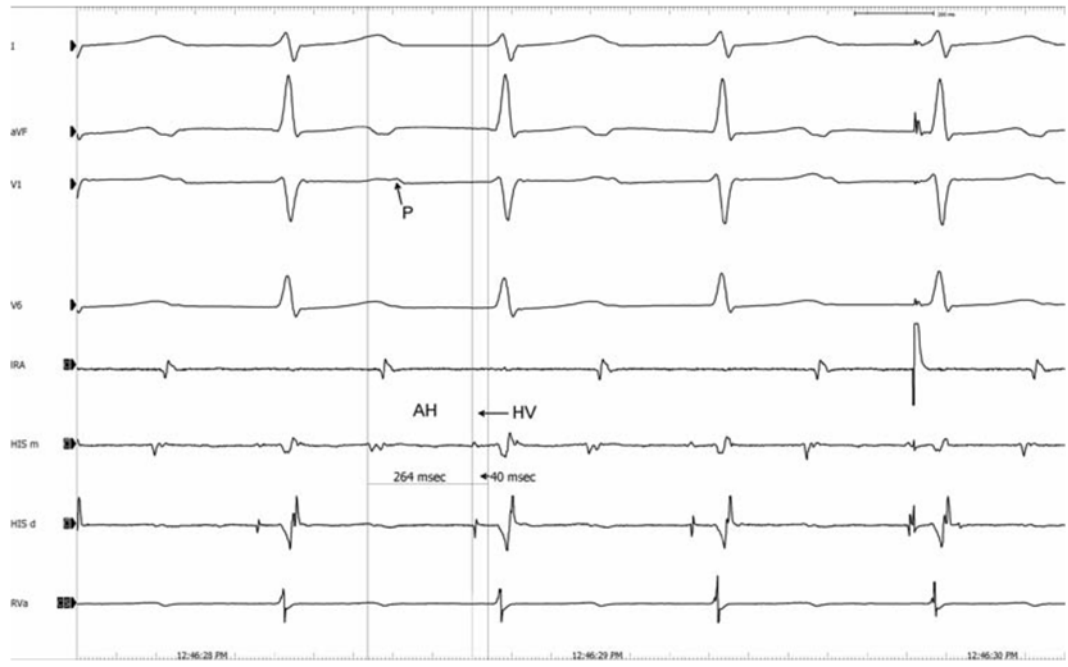

Fig. 15.13 Intracardiac study of a patient with first-degree AV block. Note prolonged AH and normal HV intervals localizing conduction delay to the AV node

primarily guide diagnosis and management of first- and second-degree AV block. Electrophysiologic study is not routinely performed solely for the assessment of first- and second-degree AV block because surface electrocardiograms provide adequate information for management. Invasive electrophysiologic studies of AV node function can be performed as an adjunct to hemodynamic study when deemed useful.

Measurements relevant to AV conduction include the intra-atrial conduction time (from the high right atrium near the sinus node to the low septal right atrium near the AV node), the AH interval, a measure of AV nodal conduction, and the HV interval, which reflects conduction through the His–Purkinje system to the ventricles (See Chap. 3). Intracardiac recordings allow distinction of the level at which block occurs (Figs. 15.13 and 15.14). Slow intra-atrial or AV nodal conduction is almost always the mechanism for first-degree AV block and is confirmed by recording a prolonged intra-atrial conduction time or AH interval. Similarly, Mobitz I block is almost always within the AV node such that the AH interval prolongs and there is no His bundle deflection with the onset of AV block. In contrast, Mobitz II block is more frequently infranodal. The AH interval typically remains constant and there is a His bundle deflection resulting in a non-conducted impulse and no ventricular activation.

Patients with first-degree or Mobitz I second-degree AV block often have prolonged effective and functional refractory periods for the atrium or the AV node. Normally, the effective refractory period of the His–Purkinje system and ventricular tissue is shorter than the functional refractory period for the AV node. Thus, block distal to the His during programmed stimulation is abnormal, though exceeding rare in the young.

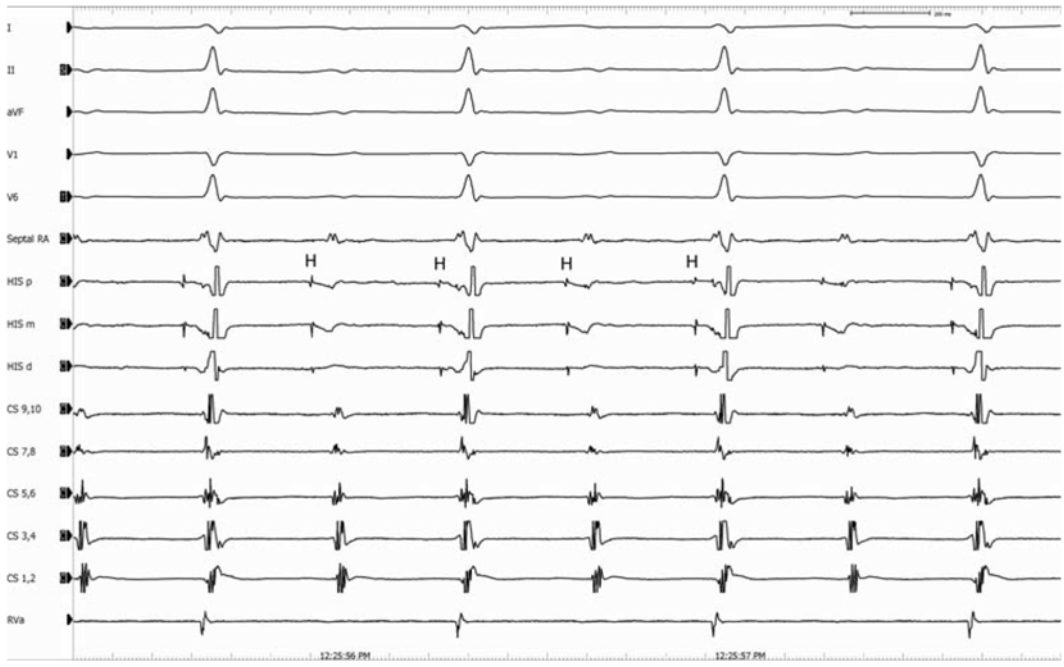

Fig. 15.14 Supraventricular tachycardia with 2:1 block. Note His depolarization (H) for conducted and blocked impulses localizing block to the distal conduction system

Prognosis and Treatment

Most children with first- and second-degree AV block do not experience progression to complete heart block and most do not require treatment. Transient first- and second-degree AV block may be observed during Holter recordings of healthy children and adolescents, particularly those who are athletes and particularly at night with predominantly sympathetic withdrawal. Avoidance of medications known to slow AV conduction is prudent, especially in those patients at risk for AV conduction system disease. For patients who are already receiving these medications, the potential benefit must be weighed against the risk of impaired conduction.

Patients who are acutely symptomatic with second-degree AV block are uncommon, but can be treated with atropine, isoproterenol, and temporary pacing. With some infectious diseases, such as Lyme carditis, the block may resolve entirely after antibiotic therapy. There are no chronic medical management options for patients with significant AV block. Consensus guidelines exist only for advanced AV block, long QT syndrome with 2:1 block or greater, and progressive AV block related to neuromuscular disease (see Chap. 16, Table 16.2). Patients with associated structural heart disease, a family history of progressive AV block or sudden death, and those known to carry the mutation of *NKX2.5* gene, require close surveillance and possibly permanent pacing as well.

Suggested Reading

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