

# Acquired Complete Atrioventricular Block in Young People: An Overview

## Bloc atrioventriculaire complet acquis chez les jeunes : Une Mise au point

Mariam Jabeur, Selma Charfeddine, Amine Bahloul, Tarek Ellouze, Faten Triki, Rania Gargouri, Leila Abid.

*HEDI CHAKER University Hospital of SFAX, Department of Cardiology*

### RÉSUMÉ

Le bloc auriculo-ventriculaire complet acquis constitue un enjeu clinique important, en particulier lorsqu'il se manifeste chez les individus jeunes. Cette mise au point vise à évaluer les étiologies, les approches diagnostiques et les stratégies de prise en charge du bloc auriculo-ventriculaire complet acquis chez la population plus jeune, en s'appuyant sur la littérature actuelle et les connaissances cliniques. Le champ d'application englobe un examen détaillé des causes sous-jacentes, allant des malformations cardiaques congénitales et des affections inflammatoires aux complications médicamenteuses et post-chirurgicales.

### MOTS-CLÉS

Bloc AV acquis, adultes jeunes, Troubles du système de conduction, Stimulation

### SUMMARY

Acquired complete atrioventricular block, a condition characterized by the complete cessation of electrical impulse transmission from the atria to the ventricles, is a significant clinical concern, particularly when it manifests in young individuals. This overview aims to comprehensively evaluate the etiologies, diagnostic approaches, and management strategies for acquired complete atrioventricular block in the younger population, drawing upon current literature and clinical insights. The scope encompasses a detailed examination of the underlying causes, ranging from congenital heart defects and inflammatory conditions to drug-induced and postsurgical complications.

### KEYWORDS

Acquired AV block, Young adults, Conduction system disorders, Pacing.

### Correspondance

Mariam Jabeur, MD  
Department of Cardiology, HEDI CHAKER University Hospital, Sfax  
Mariemjabeur2@gmail.com

## INTRODUCTION-OBJECTIF

Acquired complete atrioventricular block, a condition characterized by the complete interruption of electrical impulse transmission from the atria to the ventricles, represents a significant clinical challenge, particularly when it occurs in young individuals. In complete heart block, no atrial impulses reach the ventricles [1]. The causes, clinical significance, and management strategies for acquired complete atrioventricular block in this population require careful consideration. This review aims to comprehensively evaluate acquired complete atrioventricular block in young people, encompassing its etiology, diagnostic approaches, and therapeutic interventions.

Acquired complete atrioventricular block in young people can arise from a multitude of factors, distinguishing it from congenital etiologies or age-related degenerative processes more commonly observed in older populations.

## ETIOLOGY AND PATHOPHYSIOLOGY

The etiology of acquired complete atrioventricular block in young individuals is diverse, encompassing congenital heart defects, surgical interventions, inflammatory conditions, drug-induced causes, and, in rare instances, idiopathic factors. Congenital heart defects, particularly those necessitating surgical repair, can predispose young patients to the development of atrioventricular block [2]. Surgical interventions, while often life-saving, carry the inherent risk of iatrogenic injury to the conduction system [3]. The atrioventricular node, situated anteriorly in the right atrium, is vulnerable to fibrosis with advancing age, making patients prone to developing atrioventricular block [2]. The risk of sinus node dysfunction and complete heart block requiring permanent pacing increases with age [2]. Specifically, surgical closure of atrial septal defects has been associated with an increased risk of late-onset complete heart block, potentially manifesting years after the initial corrective procedure [2]. Inflammatory conditions, such as acute rheumatic fever, Lyme disease, and myocarditis, can induce inflammation and subsequent damage to the conduction system, leading to atrioventricular block.

The etiology of acquired complete atrioventricular block in young individuals is diverse, encompassing inflammatory, infectious, iatrogenic, and drug-induced causes. Inflammatory conditions, such as rheumatic fever,

systemic lupus erythematosus, and sarcoidosis, can trigger atrioventricular block through direct inflammation and fibrosis of the conduction system. Infectious etiologies, including Lyme disease, can induce atrioventricular block via direct invasion of the cardiac tissue by pathogens or through immune-mediated mechanisms. Iatrogenic causes, particularly those associated with cardiac surgery for congenital heart disease, represent a significant proportion of cases in young people [4]. Inflammatory diseases can trigger conduction system disturbances and subsequent atrioventricular block through various pathophysiological mechanisms, including direct inflammatory damage, fibrosis, and immune-mediated injury to the cardiac conduction pathways.

Drug-induced atrioventricular block, while less common, can occur with medications such as beta-blockers, calcium channel blockers, and digoxin, especially in individuals with underlying conduction system abnormalities. The risk of post-tachycardia pauses and subsequent arrhythmias should be considered, especially after conversion of atrial fibrillation [5]. Understanding the specific etiology is essential for guiding appropriate management strategies and predicting long-term outcomes.

The pathophysiology of acquired complete atrioventricular block involves disruption of the normal electrical conduction pathway within the heart. This disruption can occur at the level of the atrioventricular node, the His bundle, or the bundle branches, leading to complete dissociation between atrial and ventricular activity. In cases of atrioventricular nodal block, a junctional escape rhythm typically emerges, characterized by a narrow QRS complex and a relatively stable heart rate. In contrast, block distal to the His bundle often results in a slower, more unstable ventricular escape rhythm with a wide QRS complex [3]. The precise mechanism underlying conduction system dysfunction varies depending on the underlying etiology, ranging from inflammation and fibrosis to direct cellular damage. The severity of the block and the stability of the escape rhythm determine the clinical presentation and the need for intervention.

## ETIOLOGY AND PATHOPHYSIOLOGY

The clinical presentation of acquired complete atrioventricular block in young people varies widely, ranging from asymptomatic bradycardia to syncope and sudden cardiac arrest. Symptoms such as fatigue, exercise

intolerance, dizziness, near syncope, or syncope may prompt evaluation for underlying cardiac conditions [6]. Young individuals may exhibit subtle signs of reduced cardiac output, such as decreased exercise capacity or unexplained fatigue. The severity of symptoms often depends on the underlying heart rate, the stability of the escape rhythm, and the presence of coexisting cardiac conditions. In some cases, complete atrioventricular block may be detected incidentally during routine electrocardiographic screening or evaluation for other medical conditions.

Electrocardiography is the cornerstone of diagnosis, revealing complete dissociation between atrial and ventricular activity, with the absence of any relationship between P waves and QRS complexes. The ventricular rate is typically slow, ranging from 30 to 50 beats per minute, depending on the level of the block and the inherent automaticity of the escape pacemaker. Additional diagnostic modalities, such as Holter monitoring and event recorders, may be useful in capturing intermittent or paroxysmal episodes of atrioventricular block. Electrophysiological studies may be indicated in select cases to further evaluate the level and mechanism of the block, particularly when considering interventional management strategies.

## DIAGNOSTIC EVALUATION

The diagnostic evaluation of acquired complete atrioventricular block in young people involves a multi-faceted approach, integrating clinical history, physical examination, electrocardiography, and advanced imaging techniques. Electrocardiography remains the cornerstone of diagnosis, revealing complete dissociation between atrial and ventricular activity, with the atrial rate exceeding the ventricular rate. Holter monitoring can be valuable in detecting intermittent or paroxysmal atrioventricular block, while exercise testing can assess the heart rate response to exertion and identify chronotropic incompetence. Electrophysiologic studies may be indicated in select cases to further elucidate the mechanism of the block and identify the site of conduction delay. Additionally, comprehensive laboratory testing should be performed to evaluate for underlying inflammatory, infectious, or autoimmune etiologies. Cardiac magnetic resonance imaging can provide valuable insights into the presence of myocardial inflammation, fibrosis, or structural abnormalities, aiding in the differential diagnosis of acquired atrioventricular block [2]. If careful scanning of the cardiac cycle is performed on all patients with intermittent

bundle branch block, accessory pathways, and malfunctioning pacemakers, supernormal excitability and conduction may be unmasked [7].

Molecular screening is now part of the diagnostic process [8]. The integration of genetic testing into the diagnostic evaluation of atrioventricular block holds promise for refining risk stratification, guiding personalized management strategies, and potentially identifying novel therapeutic targets [9], [10].

## MANAGEMENT STRATEGIES

### *Caractéristiques générales de la population*

Management strategies for acquired complete atrioventricular block in young people are individualized based on the underlying etiology, symptom severity, and presence of comorbidities. In cases of reversible causes, such as drug-induced or Lyme-related atrioventricular block, the primary focus is on addressing the underlying condition, with temporary pacing potentially employed as a bridge to recovery. However, in cases of irreversible or high-grade atrioventricular block, permanent pacemaker implantation is generally indicated to restore physiological heart rates and improve cardiac function. Conventional transvenous pacing has been the mainstay of treatment for decades, but it is associated with potential long-term complications, such as lead dislodgement, infection, and venous obstruction. Recent advances in pacing technology, such as leadless pacemakers and conduction system pacing, offer promising alternatives to conventional transvenous pacing, potentially minimizing the risk of lead-related complications and optimizing ventricular synchrony. In adults with congenital heart disease, permanent pacing is recommended for symptomatic sinus node dysfunction or symptomatic bradycardia with any degree of atrioventricular block [2]. His bundle or left bundle branch pacing was attempted in 15 CCTGA patients [3]. With CCTGA, the anomalous His bundle course can make conventional cardiac resynchronization therapy problematic.

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Conduction system pacing can be achieved by either His bundle pacing or proximal left bundle branch pacing [3]. His bundle pacing has been attempted in patients with congenitally corrected transposition of the great arteries [3]. It's important to note that the decision to proceed with permanent pacing should be individualized based on the patient's specific clinical circumstances and preferences. Temporary pacing may be warranted in the acute setting, such as during an episode of symptomatic bradycardia or while awaiting permanent pacemaker implantation.

Left ventricular septal pacing can reduce ventricular dyssynchrony and improve cardiac function relative to right ventricular apex pacing [11]. Ventricular pacing with a long AV delay and low backup rate are usually sufficient [2]. However, if there is poor AV conduction, it may be preferable to use a shorter AV delay to improve AV synchrony and accept a ventricular pacing instead [2]. VVI pacing should be avoided in the absence of atrial tachyarrhythmias, since this programming may result in loss of AV synchrony [2].

In patients with CCTGA, ventricular pacing can lead to right ventricle failure [3]. Cardiac resynchronization therapy has been proposed to improve systemic right ventricular function and heart failure symptoms in SRV patients [2]. CRT may be useful in patients with SRV ejection fraction  $\leq 35\%$ , New York Heart Association function class II or above and wide QRS complex  $> 150$  ms with a complete right BBB QRS morphology [2]. However, indications for CRT are not well defined and its potential benefit remains uncertain [2]. It's still uncertain in the long term whether CRT is helpful or not [2].

ICD implantation may be considered in select cases of acquired complete atrioventricular block with an increased risk of sudden cardiac death [2]. After evaluation to define the cause of the event and exclude any reversible causes, ICD implantation is recommended for patients with CHD who are survivors of an aborted cardiac arrest [2]. Device-related infections are a common late complication, which can affect the pocket into which the generator is placed or the leads themselves [2].

The implantation of cardiac devices in patients with systemic right ventricles should ideally be performed in specialized centers by trained electrophysiologists [2]. Patients require ongoing monitoring and follow-

up to assess device function, optimize programming, and detect any potential complications. Vascular access may be challenging in SRV, and understanding cardiovascular anatomy is crucial [2].

Tertiary expertise and future collaborative research are needed to further the care for this challenging patient population [2].

## FUTURE DIRECTIONS

Further research is needed to identify novel biomarkers for predicting the development and progression of acquired complete atrioventricular block in young people. In addition, well-designed clinical trials are warranted to compare the long-term efficacy and safety of different pacing modalities in this population, including leadless pacemakers and conduction system pacing. These include establishing standardized protocols for evaluating the need for pacing in children and young adults with congenital heart disease [2]. Further studies are also needed to assess the impact of acquired complete atrioventricular block on long-term cardiovascular outcomes, including the risk of heart failure, sudden cardiac death, and overall mortality. Long-term outcomes in patients with new permanent pacemaker implantation following transcatheter aortic valve replacement have been studied, and cusp-overlap view reduces conduction disturbances and permanent pacemaker implantation after transcatheter aortic valve replacement even with balloon-expandable and mechanically-expandable heart valves [12]. His bundle pacing has emerged as a more attractive alternative to biventricular pacing for cardiac resynchronization therapy, stimulating the ventricles through the intrinsic His-Purkinje system and resulting in synchronous and more physiological electrical and mechanical activation [13], [3].

## CONCLUSION

Acquired complete atrioventricular block in young people represents a complex and heterogeneous condition with diverse etiologies, clinical presentations, and management

strategies. A comprehensive approach to diagnosis and management is essential to optimize outcomes and improve the quality of life for affected individuals. The awareness of the detrimental effects of arrhythmias on any underlying cardiac substrate has led to stronger efforts in developing novel methods and approaches to treat arrhythmia and improve patients' health and quality of life [14]. New technologies may become feasible for CHD, thus avoiding issues related to traditional device implantation [2].

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