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Voltage mapping and right ventriculography to guide ablation for arrhythmogenic right ventricular cardiomyopathy ventricular tachycardia: a case report

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Abstract
Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a family inherited cardiomyopathy associated with ventricular arrhythmias. With the development of molecular biology, histology, imaging, and other diagnostic techniques, the diagnosis rate and incidence of ARVC have gradually increased. However, ARVC remains rare in clinical practice. Currently, the diagnosis and management of ARVC is far from satisfactory in clinical practice. In the case report, we described a clinical case of radiofrequency ablation guided by voltage mapping and right ventriculography in the treatment of ARVC with ventricular tachycardia and discussed the relevant literatures.

Keywords Arrhythmic right ventricular cardiomyopathy, Ventricular tachycardia, Radiofrequency ablation, Voltage mapping, Right ventriculography

Introduction

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a rare inherited cardiomyopathy, also known as arrhythmic right ventricular dysplasia [1]. It is one of the leading causes of sudden cardiac death(20% of deaths) in young people and athletes according to an Italian study [2]. Therefore, early diagnosis and treatment of ARVC are crucial. However, ARVC was easily missed due to the mild structural abnormalities or even normal in the early clinical stage. Currently, definitive diagnosis of ARVC has been relatively complicated. The current clinical treatment for ARVC is palliative, of which the main purpose is to relieve the symptoms of arrhythmia and heart failure to reduce the risk of sudden cardiac death while improving life quality. Although some researches had indicated that catheter ablation of ventricular arrhythmias was an important therapeutic option for patients with ARVC, particularly in young patients [3, 4], the strategies for ablation and efficacy of catheter ablation were not investigated comprehensively. Here, we described the diagnosis and management of a patient with ARVC complicated by ventricular arrhythmias and successful ablation significantly alleviated ventricular tachycardia, guided by a novel method combined use of voltage mapping and right ventriculography. During more than 2 years follow-up, the patient's condition is still stable.

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Case presentation

A 30-year-old man came to our hospital on January 10, 2022, complaining of sudden palpitations. The emergency electrocardiogram (ECG) showed ventricular tachycardia (VT) which converted to sinus rhythm after intravenous amiodarone. Dynamic ECG showed frequent premature ventricular contractions (PVC), multifocal nonsustained ventricular tachycardia (NSVT). Echocardiography showed normal size of each chamber, mild tricuspid regurgitation, normal ejection fraction (63%). Normal blood routine, electrolytes, and thyroid function were investigated. The patient had no symptoms of heart failure, such as chest tightness, shortness of breath, or fatigue. Based on the results of laboratory tests and echocardiography, the patient was temporarily given metoprolol succinate 47.5 mg p.o. qd without /anti-heart failure treatment. The patient still had PVC and NSVT. Therefore, we planned to perform radiofrequency ablation. The ECG before surgery showed three forms of PVC, of which lead V1 and V2were both"QS" type. It was considered that all three PVCs might originate from the right ventricle (Fig. 1A). Leads II, III, and aVF of PVC1 were "Rs" type, which suggested that PVC1 originated from the anterior wall of the tricuspid valve annulus. However, leads II, III, and aVF were "QS" type, and lead aVR was "r" and "QrS" type respectively in PVC2 and PVC3, which suggested that these two PVCs originated from the posterior inferior wall of the right ventricle near apex and the posterior wall of the tricuspid annulus respectively. Intraoperative VT was induced, with "QS" type of leads II, III, aVF, and anterior chest leads, as well as"R" type of lead aVR (Fig. 1B). Different from the three types of PVC before surgery, it was considered that the intraoperative VT originated from the apex of the right ventricle. According to the abnormal performance of ECG, the possibility of right ventricular cardiomyopathy was considered. Unfortunately, there was no obvious abnormality in the patient's echocardiography, and we did not perform cardiac magnetic resonance imaging before the radiofrequency ablation.

We performed right ventricular angiography for further diagnose, which showed aneurysm with significantly weakened myocardial contraction at the apex of the right ventricle (Fig. 2A), suggesting the presence of myocardial fibrosis and scar at the region. Likewise, right ventricular substrate mapping under sinus rhythm showed obvious low voltages in the right ventricular apex aneurysm and its surrounding myocardium (Fig. 2B), which was a further confirmation of cardiomyopathy at the right apex. In addition, late potential mapping under sinus rhythm also showed a large number of low-amplitude fragmented late potentials in the right ventricular apical aneurysm, the latest excitation located in the aneurysm, and slow conduction in the myocardium around it (Fig. 3). Due to the

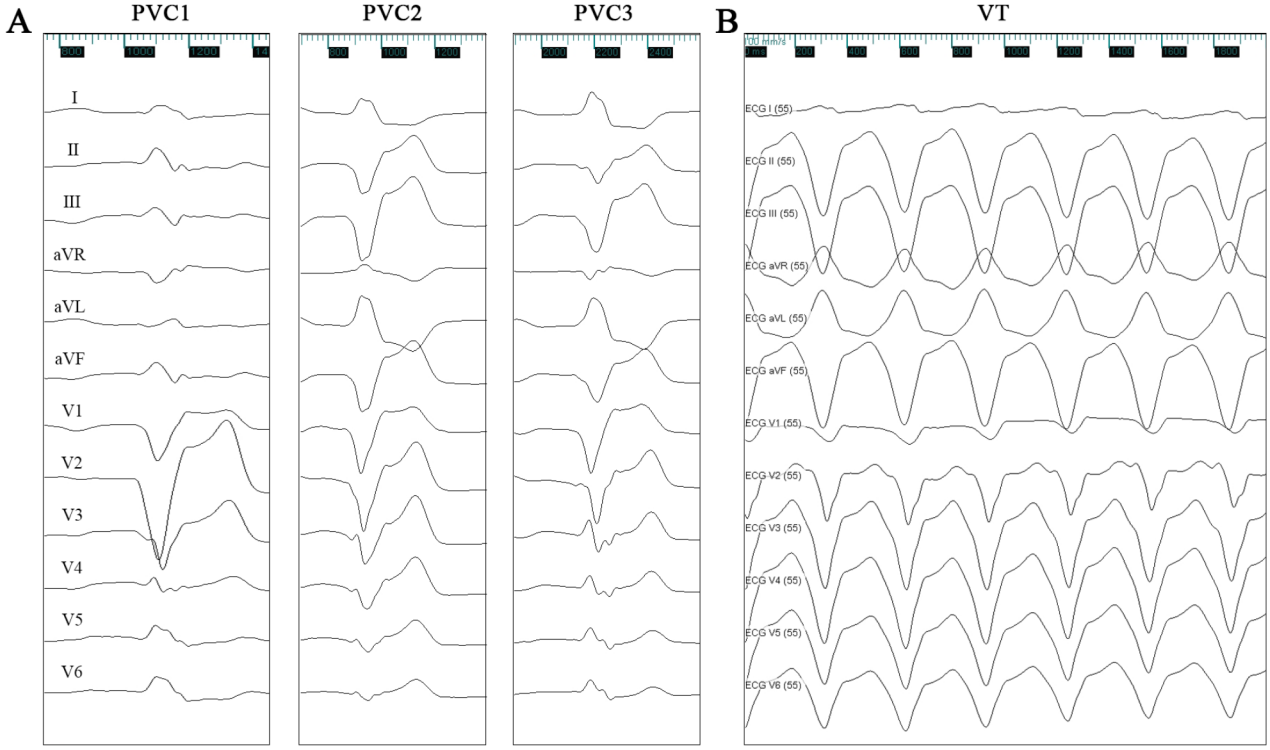

Fig. 1 Preoperative and intraoperative twelve-lead surface electrocardiogram. (**A**). Three different forms of PVC before operation; (**B**). Electrocardiogram during operation showed VT

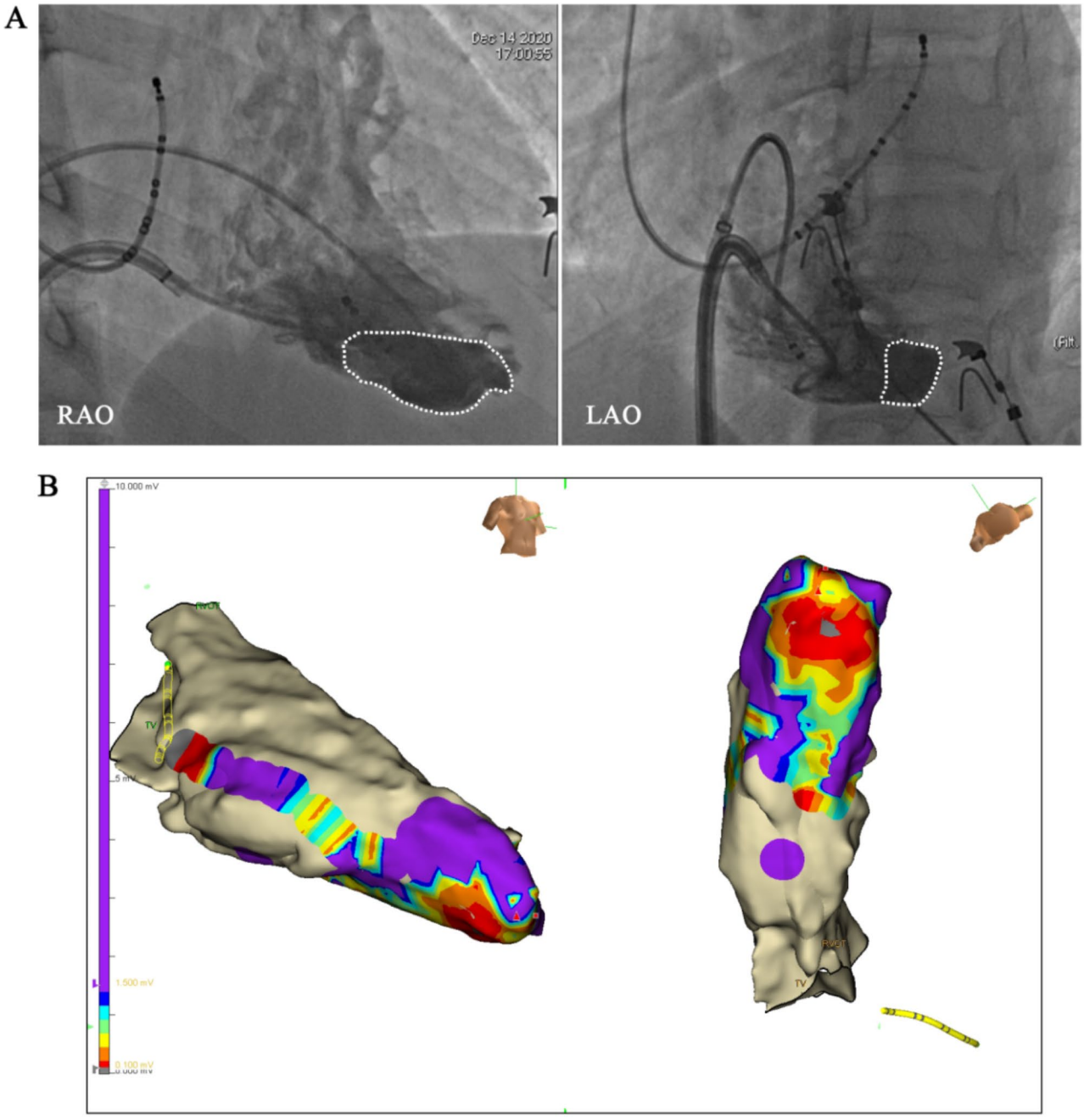

Fig. 2 Right ventriculography and voltage matrix mapping in sinus rhythm. (**A**). Right ventricular angiography shows the formation of right ventricular apical aneurysm, and the local systolic motion was weakened (dotted line); (**B**). Voltage matrix mapping under sinus rhythm showed a large area of low voltage in the right ventricular apical aneurysm

induction of NSVT and hemodynamic instability during the operation, homogenization ablation was performed at the abnormal late and low potential area of the right ventricular apex (Fig. 4A). NSVT of a morphology consistent with the clinical VT (Fig. 4B) was induced during the homogenization ablation. After the ablation, PVC2 and PVC3 disappeared, and VT was no longer induced by intravenous infusion of isoproterenol and repeated ventricular stimulation.

In addition, because of rae occurrence of PVC1, ablation was terminated. Based on the patient's medical history, VT and PVC morphology in ECG, the results of right ventricular angiography and the intraoperative voltage matrix mapping, the patient was diagnosed as ARVC. The results of comprehensive genetic analysis also further

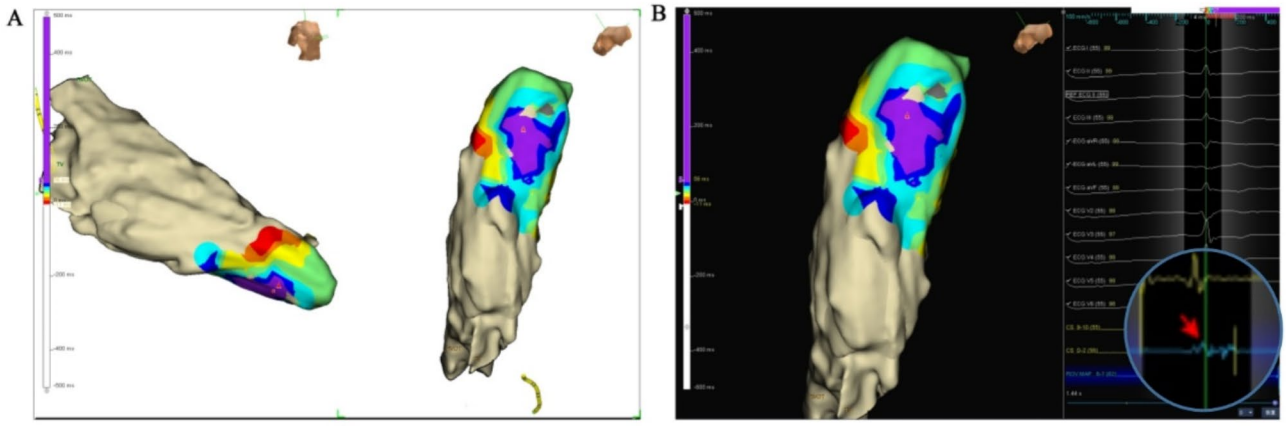

Fig. 3 Late potential mapping results of right ventricular apical aneurysms in sinus rhythm. (**A**). The late potential mapping results indicated that the latest activation was located in the aneurysm; (**B**). A large number of low-amplitude fragmented late potentials were recorded in the aneurysm

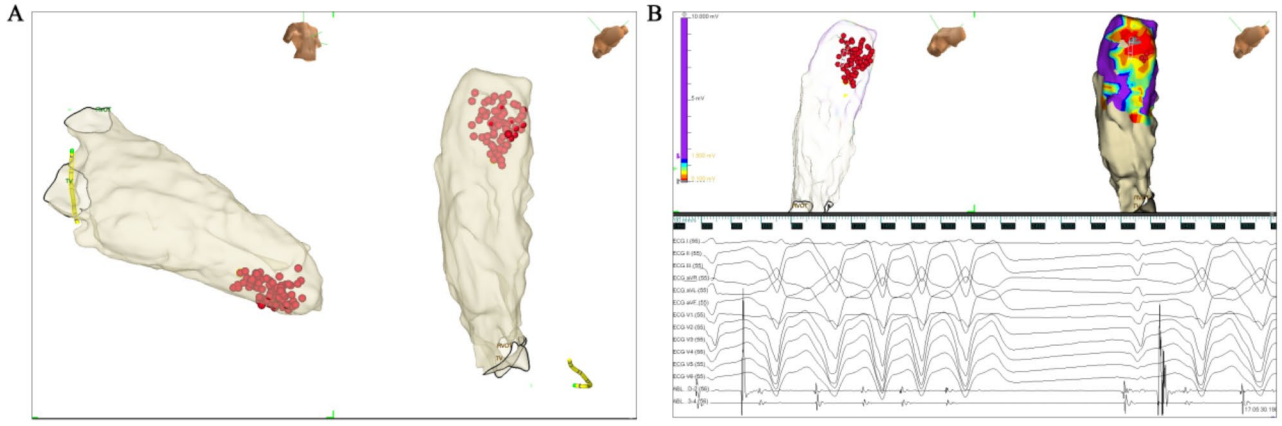

Fig. 4 Ablation of voltage matrix homogenization associated with right ventricular apical aneurysm. (**A**). Homogenized ablation point in the low-voltage area of ventricular aneurysm(saline perfusion mode: power 35 W, maximum temperature 43℃, flow rate 17 ml/min); (**B**). Short-term paroxysmal VT stimulated during ablation

Table 1 Genetic test results, according to the genetic test results, the patient was consistent with ARVC gene alteration

Gene	The reference sequence	Position of chromosome	Nucleotide alteration	Amino acid alteration	Heterozyg-osity	Pathogeni-city	Varia-tion pro-sourc-e
Pkp2	NM_004572	chr12:32949042	c.2489 + 1G > A	-	hybrid	Possible pathogenic	

Related disease information of the mutated gene

Gene	Related diseases	Genetic way
Pkp2	Arrhythmogenic right ventricular dysplasia type 9 (OMIM 69040)	Autosomal dominant inheritance(AD)

confirmed the patient's diagnosis (Table 1). There was no recurrence of VT during the next 2 years' follow-up. The re-examination of the dynamic ECG showed a significant decrease in PVCs, and NSVT was rarely detected.

Discussion

ARVC is an autosomal hereditary cardiomyopathy. The age of onset is usually between 20 and 40 years old. The overall incidence is about 0.02%, and the incidence in Europe and the United States can reach 0.05% [2, 4]. The disease was originally thought to be a congenital defect of right ventricular myocardial dysplasia. After the discovery of a genetic defect in the cardiac desmosomes, ARVC was defined as a cardiomyopathy. Subsequently, ARVC was considerded as a type of cardiomyopathy by the American Heart Association [5]. Clinical manifestations include the early concealed stage, the electrical change stage, and the structural change stage. Palpitations,

syncope, and even sudden cardiac death due to fatal arrhythmias and slowly progressive ventricular dysfunction are common. Pathological features are progressive loss of right ventricular.

myocardium and replacement by fibro-fatty tissue. The features of ECG and echocardiography are ventricular arrhythmia and structural abnormalities of the right ventricle respectively. ARVC is a progressive disease, the early cardiac structure changes are not obvious or localized in the right ventricle, and the left ventricle may be involved in the late stage. The most commonly involved sites are the anterior wall of the conus arteriosus, the apex of the right ventricle, and the subbasal side of the right ventricular tricuspid valve, forming the so-called "triangle of dysplasia" which is considered a hallmark of ARVC [6]. At present, most of the mutations are believed to be located in desmosomal coding genes, mainly PKP2 [1, 6].

The ECG of the patient in the case showed VT with a left bundle-branch block pattern. Right ventricular angiography showed the formation of right ventricular apical aneurysm, and weakened myocardial motion. Voltage matrix mapping confirmed obvious myocardial fibrosis in the right ventricular apical aneurysm. Our case met the three main diagnostic criteria, therefore, the diagnosis of ARVC was confirmed. Because the early structural changes of ARVC were not obvious or localized to the right ventricle, conventional echocardiography was difficult to detect the changes, and patients with early ARVC were easily missed. At present, cardiac MRI is also an important and effective technique for diagnosis of cardiomyopathy. Nevertheless, cardiac MRI is not a routine preoperative examination because of its time consuming and complicated examination process.

We performed right ventricular angiography combined with matrix mapping during the procedure, and the radiofrequency ablation achieved a satisfactory result by homogenization ablation in the case. Previous studies have shown that image integration combined with voltage mapping improved the success rate of radiofrequency ablation [7, 8]. However, cardiac MRI cannot observe synchronously during the operation, and it cannot be used for patients with implanted electrical devices incompatible with MRI. Cardiac MRI is not a routine examination for ARVC patients without obvious cardiac structural changes in the early stage. Therefore, right ventricular angiography combined with voltage mapping may be a better choice for those patients.

The clinical manifestations such as malignant arrhythmia and heart failure, are controlled by medicines and devices, which is a main treatment target of ARVC. Treatment strategies include class I, II, III antiarrhythmic drugs, ICD implantation, and catheter ablation [6, 9]. Unfortunately, there is no specific drug to control or eliminate arrhythmia, or to change the natural course of the disease, and no drug alone or in combination can completely prevent sudden death for ARVC patients. A study by Orgeron GM et al. showed that ICD implantation is a safe and effective measure to improve the natural history of ARVC and prevent sudden death [2]. However, ICD implantation cannot control the onset of malignant arrhythmias, and it may lead to unwanted results, such as pain, anxiety, restlessness when patients are treated by electrical shock during ventricular tachycardia storms repeatedly. Therefore, antiarrhythmic drugs should be used as adjuvant therapy for ARVC patients with frequent ICD discharges. Catheter ablation is currently the most effective method to reduce or even eliminate VT [4, 10]. Recent evidence-based medical results from the largest number of cases in Asia on ARVC catheter ablation showed that both transendocardial and transepicardial catheter ablation could reduce the burden of VT episodes in ARVC patients, and even achieved clinical cure for a long time, the effective rate was about 71% [11, 12]. The patient in our case got VT as the main clinical manifestation. The latest research showed, long-term VT-free survival had been achieved in over half of ARVC patients following ENDO-only-ablation, increasing to over 75% if VT noninducibility was achieved [11]. In our case, we also achieved a better clinical outcome with only epithelial ablation.

Echocardiography showed no obvious structural changes of the right ventricle. Intraoperative right ventricular angiography and voltage mapping confirmed that the cardiomyopathy was limited to the apex of the right ventricle. Considering that the origin of VT was highly consistent with the location of myocardial lesion, catheter ablation would probably eliminate the VT. In fact, VT was no longer induced after the homogenizational ablation of the diseased myocardium, and no VT occurred during postoperative follow-up. Although the patient had no clear family history, early ICD implantation was still recommended for him in consideration of the particularity of ARVC and ICD implantation indications, the patients refused it. Thus, regular follow-up was arranged for him.

Conclusion

This case had important clinical implications. First of all, the possibility of early manifestations of ARVC should be ruled out in patients with multifocal PVCs originating from the non-outflow tract of the right ventricle, especially when the morphology of the ventricular tachycardia was not consistent with ventricular premature contraction. Secondly, echocardiography often failed to detect early structural changes in ARVC. Therefore, it was necessary to complete ventriculography or cardiac MRI to determine whether the myocardial structure was

abnormal. Finally, the combined application of voltage mapping and right ventriculography improved satisfactorily the efficacy of radiofrequency catheter ablation of VT originated from right ventricle.

Abbreviations

VT	Ventricular tachycardia
ARVC	Arrhythmogenic right ventricular cardiomyopathy
ECG	Electrocardiogram
PVC	Premature ventricular contraction
ICD	Implantable cardioverter-defibrillators

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Author contributions
Xinrong Fan and Chengying Yang designed the review and wrote the manuscript. Yihua Cai, Yan Wei & Gang Li also have participated sufficiently in the work to take public responsibility for appropriate por-tions of the content and agreed to be accountable for all as-pects of the work in ensuring that questions related to its accuracy.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
Not applicable.

Competing interests
The authors declare no competing interests.

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