ARRHYTHMIAS IN CHILDREN AND ADULTS WITH CONGENITAL HEART DISEASE

KEY POINTS:

1. The general principles of arrhythmia care in children are similar to those of adult care, including the essential clinical evidence of an ECG recorded during symptoms, although children are less able to report their symptoms, and some arrhythmias in children can result in tachycardiomyopathy and heart failure.
2. The unborn child can be affected by life-threatening arrhythmias.
3. There are a number of arrhythmias in children that do not occur in adults.
4. Children with no structural heart disease are suitable for curative catheter ablation, but the timing of this requires careful judgement because of the risk of complications from catheters and devices designed for adult care, and because ablative lesions may grow with the child and future safety is still uncertain.
5. Children and adults with congenital heart disease commonly have complex arrhythmia problems, and these need to be managed in multi-disciplinary teams of paediatric cardiologists, electrophysiologists, experienced paediatric surgeons and support staff, not small local departments.
6. Children and adults with congenital heart disease and arrhythmias may need hybrid therapy with antiarrhythmic drugs, pacemakers and ICDs, catheter ablation and surgery. Such complexity argues for these services to be provided only in tertiary centres undertaking a large amount of such work and providing adequate expertise and resources.
7. Centres treating arrhythmias in children and patients with congenital heart disease should have close links with clinical genetics services.
8. Training programmes for paediatric cardiac electrophysiology should be established.

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INTRODUCTION

The main aim of the chapter is to highlight clinical, technical and managerial issues associated with cardiac arrhythmias that present in childhood. There are important differences in clinical presentation and management of the child in comparison to the adult. There are also unique and complex problems associated with cardiac arrhythmias in the adult with congenital heart disease. Recommendations are made on the appropriate models of care and provisions of services necessary for this important group of patients.

ARRHYTHMIAS IN THE CHILD

Arrhythmias can present at any time in childhood from foetal life to adolescence. The nature of presentation varies according to the age of the child. A classification of arrhythmias in children is shown in Figure 1.

Many paediatric arrhythmias have a similar electrophysiological mechanism to those found in adults. There are, however, certain cardiac arrhythmias that are unique to children and either not seen in adulthood or behave very differently to the adult equivalent. Examples of these are shown in Figure 2. In particular, certain tachycardias that present in infancy may be refractory to medical treatment requiring several antiarrhythmic drugs. Despite this, many will resolve completely after 12-18 months as a consequence of myocardial maturation and growth. A detailed description of individual arrhythmias is beyond the scope of this chapter but is available in the scientific literature.[1 2]

Figure 1. Classification of arrhythmias in children

Structurally normal heart	Bradycardia	Heart block
		Sinus node dysfunction
	Tachycardia	Supraventricular tachycardia
		Ventricular tachycardia
Structurally abnormal heart	Bradycardia	Heart block
		Sinus node dysfunction
	Tachycardia	Supraventricular tachycardia
		Ventricular tachycardia

In comparison to adults, children have poor and sometimes absent symptom recognition. Consequently, they may present late with a cardiomyopathy secondary to chronic tachycardia ("tachycardiomyopathy") or may be extensively investigated for non-specific symptoms by general paediatricians or paediatric neurologists. Similarly, the response of a young child to antiarrhythmic drugs – especially in infancy- may differ considerably from an adult. Finally, catheter ablation techniques and implantable devices available for the treatment of cardiac arrhythmias are invariably designed for adults.

Figure 2. Paediatric arrhythmias seldom seen in adults

Arrhythmia	Presentation	Treatment	Outcome
Permanent Junctional Reciprocating Tachycardia	Infancy – school age	medication often unhelpful Radiofrequency ablation	Cardiomyopathy unless treated
Neonatal atrial Flutter	Neonate	DC cardioversion antiarrhythmics	resolves
Junctional ectopic Tachycardia	Infancy – teens	Antiarrhythmics Radiofrequency ablation	may cause cardiomyopathy
Incessant ventricular Tachycardia of infancy	Infancy	Antiarrhythmics Rarely, surgery	usually resolve
Paroxysmal Catecholaminergic Ventricular tachycardia	early/mid childhood	Beta blockers defibrillator	persists to adult life

ARRHYTHMIAS IN THE FOETUS

Detection and management of a foetal arrhythmia requires close collaboration between the foetal medicine specialist, foetal cardiologist, maternal medicine specialist, neonatologist and paediatric electrophysiologist. At present there is no reliable technique for continuous ambulatory monitoring of foetal arrhythmias. Thus, the foetus with an arrhythmia requires frequent multidisciplinary follow-up, which includes foetal echocardiography.

Foetal tachycardia may be noted as an incidental finding during routine assessment or may present with foetal heart failure (hydrops foetalis) or stillbirth. Foetal supraventricular tachycardias can be treated by administration of antiarrhythmic drugs to the mother. This is successful in approximately 90% but the recurrence risk in post natal life is relatively high with a foetal and infant mortality rate of 14% [3]. Foetal ventricular tachycardia is extremely rare and may be a feature of Long QT syndrome. Occasionally, Long QT syndrome presents with foetal heart block.

Foetal heart block affects 1:20,000 live births. Thus, around 30 infants will be born with congenital heart block each year in the United Kingdom. Of these 30-50% will have a structurally abnormal heart. Of the rest, a high proportion are associated with maternal connective tissue disease or anti-Ro and anti-La antibodies. [4] Foetal heart block may be asymptomatic and detected on a routine foetal ultrasound scan or may be associated with severe cardiac compromise. Intrauterine treatment (including foetal pacing) has proved disappointing. The mortality in childhood is high (14%) and approximately 50% require pacing in the neonatal period.[5]. Neonatal pacemakers make be implanted using the epicardial or endocardial route. With improvements in epicardial steroid eluting leads, the epicardial pacing technique is now the first choice for infants in many units[6]. Endocardial systems, however, can be implanted successfully with excellent long-term results.

Management of a foetal arrhythmia involves multidisciplinary input with subspecialised expertise and facilities including foetal and maternal echocardiography, neonatal and paediatric intensive care and cardiac surgery. It is important to keep the baby in the womb if possible. The decision on the optimal time to deliver the foetus is often extremely difficult and properly planned research

is needed to determine the best time. A lack of sufficient neonatal intensive care cots, difficulties in funding dedicated foetal and maternal echocardiography equipment in the maternity unit and lack of trained foetal cardiologists prevent good outcomes in the UK. The development of clinical networks with high quality telemedicine links is desirable to allow tertiary centres to link with the very few foetal cardiologists.

ARRHYTHMIAS IN THE CHILD - structurally normal heart

The evaluation of the child with a cardiac arrhythmia starts with a detailed clinical history and physical examination.

Questioning should:

- Ask about precipitating events.
- Be aware of age-specific presentations. (Figure 3).
- A family history of cardiac arrhythmias or unexpected sudden cardiac death (sudden arrhythmic death syndrome) suggest an inherited disorders such as arrhythmogenic right ventricular cardiomyopathy, hypertrophic cardiomyopathy or long QT syndrome.

Examination should emphasise:

- A full clinical examination.
- The presence of dysmorphic features (for example; Down's syndrome or chromosome 22q11 microdeletion facies) which may suggest underlying structural cardiac disorder.
- Non-cardiac anomalies such as sensorineural deafness may alert the clinician to the corresponding cardiac diagnosis (long QT syndrome).
- Structural cardiac defects that are easy to miss on clinical examination (eg mitral valve prolapse, atrial septal defect).
- Signs of haemodynamic compromise.

Figure 3. Age specific presentations of cardiac arrhythmias

Foetus • Antenatal tachycardia on screening • Hydrops foetalis • Neonatal heart failure.
Infant • Tachycardia on routine assessment • Signs of heart failure (poor feeding, tachypnoea) • Cardiovascular collapse • Breath holding attacks, apnoea **Young child (preschool)** • Non-specific aches and pains (e.g. tummy pain) • Vivid descriptions ("a butterfly trying to fly out my chest"). • Unexplained episodic pallor, sweating and lethargy • Faints or seizures • Breath holding attacks
Older child

- Palpitations
- Chest pain
- Faints or seizures
- Collapse and sudden death

In the well child, however, clinical examination is often unhelpful and never diagnostic. Investigation includes:

- Investigating for electrolyte imbalance, drug toxicity or poisoning in acute arrhythmias.
- Electrocardiogram (ECG).
- Echocardiogram.

Supraventricular Tachycardia

Supraventricular tachycardia (SVT) accounts for 95% of tachyarrhythmias in children. Most children with SVT have a structurally normal heart. SVT should be considered in the diagnosis of the collapsed infant even if the rhythm is sinus at presentation[7]. Cardiovascular collapse can also be the first presenting symptom in older children with Wolff-Parkinson-White syndrome but this is rare.[8 9] In other children with SVT, sudden cardiac death is extremely rare. Incessant tachycardias (for example, permanent junctional reciprocating tachycardia) usually present with symptoms of heart failure rather than palpitations.

Supraventricular tachycardia in young children is usually due to orthodromic AV re-entry tachycardia (AVRT). If this accessory pathway allows forward conduction from atria to ventricles, the ECG will show features ventricular pre-excitation of Wolff-Parkinson-White syndrome (Figure 1). If the accessory pathway can only conduct backwards from ventricles to atria, the 12 lead ECG in sinus rhythm is normal. In orthodromic AVRT the SVT has narrow complexes. In Wolff-Parkinson-White (WPW) syndrome, the circuit can go around the other way (antidromic AVRT).

In WPW Syndrome the accessory pathway may conduct rapidly if atrial fibrillation develops, and this can degenerate into ventricular fibrillation and death. Atrial fibrillation is more common in children with WPW syndrome than in the normal population. Moreover, the first symptom in a teenager with WPW syndrome may be atrial fibrillation with antidromic AVRT. Consequently, there is a 2% incidence of sudden cardiac death in adolescents and young adults with WPW syndrome followed up over 1-15 years. [10]

Ventricular Arrhythmias

Ventricular ectopic beats are common in healthy children. Benign ventricular ectopic beats arise from a single focus and disappear on exercise. An exercise ECG stress test is valuable in assessment.

In the child, supraventricular and ventricular tachycardia may present with identical symptoms. Moreover, some children with ventricular tachycardia are asymptomatic and present with abnormal neck pulsations or on screening. Ventricular tachycardia accounts for less than 5% of tachycardias in children. Most children with ventricular tachycardia have a structurally heart abnormality although this may subtle. In the infant, up to 40% have significant associated medical disorders. (Figure 4). A benign variant of infant VT exists which is usually relatively slow, well tolerated and self-limiting.

Figure 4. Medical Disorders associated with ventricular tachycardia in infancy

- Drug toxicity
- Severe electrolyte imbalance
- Congenital long QT syndrome
- Myocarditis
- Adrenogenital syndrome
- Cardiac tumours (including microscopic hamartoma and tuberose sclerosis related rhabdomyomas)

In the toddler, accidental drug ingestion (usually digoxin or tricyclic antidepressants) should be considered and in the older child, solvent and recreational drug abuse. Some forms of VT are catecholamine dependant and brought on by emotion or exercise. Ventricular arrhythmias, which are induced by exercise usually, reflect serious underlying pathology and further investigation is mandatory.

AV Block / Bradycardia
Two main types of bradyarrhythmia occur:

- Complete AV block (CAVB)
- Sinus node dysfunction (SND).

Most CAVB is either congenital or related to previous cardiac surgery. CAVB is often well tolerated in the infant and young child. The development of symptoms (syncope, presyncope,) is an indication for pacing. SND can present with similar symptoms. In infants seizures may occur, and the bradyarrhythmia may not be recognised. A misdiagnosis may occur.

Bradycardias and tachycardias may be associated with syncope, pallor and sweating. In most children syncope is simple Reflex Syncope. If syncope occurs during exercise or if there is a known underlying arrhythmia or structural heart defect, urgent cardiac assessment is indicated.

Investigation is similar in principle to adult investigation:

- As for adults, achieve symptom/ECG correlation.
- A 24-hour heart rate recording may be diagnostic in frequent symptoms.
- Exercise ECG test can be used in older children if symptoms are exercise related.
- For children with less frequent symptoms reliable event recorders are available.
- If symptoms are very infrequent a loop recorder can be implanted subcutaneously ILR. This records the ECG continuously for up to 18 months and can be programmed to automatically detect arrhythmias.
- Head-up tilt testing may be useful. This allows a positive diagnosis to be made and the institution of targeted treatment. [211]

Figure 5. Causes of Atrioventricular Block in Children

Congenital	
Idiopathic	
Structural heart defect	Isomerism Congenitally corrected transposition
Maternal connective tissue disorder	Positive antiRo, antiLa antibodies

Long QT syndrome

Acquired	
Muscular dystrophy	Myotonic dystrophy Emery Dreifuss dystrophy Duchenne dystrophy Kearns-Sayre
Infective / Immune mediated	Rheumatic fever Lyme disease Chagas's disease Rubella HIV infection Diphtheria Myocarditis (especially mycoplasma and coxsackie)
Iatrogenic	Post cardiac surgery Post radiofrequency ablation
Other	Idiopathic Familial dysautonomia

ARRHYTHMIAS IN THE CHILD - structurally abnormal heart

Arrhythmias are common in children with congenital heart disease. Arrhythmias may occur as a consequence of the underlying structural abnormality, as a consequence of haemodynamic changes such as atrial stretch or as a result of surgical scars. Late postoperative arrhythmias are discussed in the section on the adult with congenital heart disease. These arrhythmias, however, can present in childhood and adolescence. In many patients, the development of an arrhythmia reflects an underlying haemodynamic problem. Management of an arrhythmia in the child with congenital heart disease should therefore include a detailed haemodynamic assessment. Increasingly, the best method of prevention of post surgical arrhythmias may be to consider concomitant arrhythmia and structural heart surgery using recognised principles to avoid the creation of re-entry circuits.[12-14]

Children with congenital heart disease may also have accessory pathways.[15] These may be treated with conventional radiofrequency techniques but diagnosis is facilitated by the use of electroanatomic mapping techniques.[16] Similarly, heart block can occur as part of the natural history of a congenital heart defect such as left atrial isomerism. [17]

Sudden Cardiac Death In The Child

The sudden, unexpected death of a child is a rare but devastating event. The individual age-related risk is about 1:20,000 to 1:50,000. [18] The main cardiac causes for sudden cardiac death in children are described in Figure 6.

Figure 6. Cardiac causes for sudden death in childhood (adapted Wren [18])

Cardiovascular malformations
Aortic stenosis.
Pulmonary atresia.
Coarctation of aorta.
Pulmonary vascular disease.
Coronary artery malformations.
Tetralogy of Fallot (post operative).
Transposition of great arteries (post atrial switch).

Other structural heart abnormalities
Cardiomyopathy (dilated, restrictive and hypertrophic).
Myocarditis.
Kawasaki Disease.
Arrhythmogenic right ventricular cardiomyopathy.

Primary arrhythmia
Long QT syndrome.
Short QT syndrome
Wolff Parkinson White syndrome.
Atrioventricular block.
Other ventricular arrhythmias (Catecholaminergic ventricular tachycardia and Brugada syndrome).
Non-hypertrophic variants of Hypertrophic cardiomyopathy (troponin t mutation).

Children with congenital heart disease at risk of sudden cardiac death should undergo lifelong follow up by a paediatric or congenital heart disease specialist. Data are accumulating on preventative measures for certain conditions. Thus, in Tetralogy of Fallot large multicentre studies have enabled risk stratification for malignant arrhythmias to be developed and treatment strategies proposed.[19-21] In other conditions such as hypertrophic cardiomyopathy, research carried out in families (mostly adult) has allowed an extrapolation of preventative measures and risk stratification to be applied to children.[22] In a population based survey of sudden death at age 1-20 years in the northern region of England, 11% (270) of deaths were sudden. [23] Of these, 33 were due to known pre-existing cardiovascular anomalies and a further 26 due to cardiovascular abnormalities diagnosed at autopsy. (Figure 7a 7b). Undiagnosed hypertrophic cardiomyopathy caused less than 1 death per million person years but unexplained death – which the authors suggested might be due to primary cardiac arrhythmias- was 10 times more common.

A screening programme in Japanese children has demonstrated that a screening ECG can detect children with the potential for dangerous cardiac arrhythmias (A-V block, long QT syndrome, Wolff Parkinson White syndrome).[24] However, it has not yet been shown that such a programme prevents sudden death. Recommendations on the medical approach to sudden cardiac death have been published but these are primarily designed for the adult population.[25] Similar consensus recommendations for children do not exist. Recently, recommendations on sports participation for young patients with genetic cardiovascular disease have been published.[26] These include recommendations on primary arrhythmias including arrhythmogenic right ventricular cardiomyopathy, long QT syndrome, hypertrophic cardiomyopathy, Brugada syndrome and catecholaminergic polymorphic ventricular tachycardia.

Any child suspected of having cardiac arrhythmias should have a resting 12-lead ECG with an appropriate report.

NON INVASIVE INVESTIGATIONS

A 12 lead ECG should be obtained in every child suspected of an arrhythmia but the key to diagnosis is to obtain an ECG during symptoms. Most children with an arrhythmia have a structurally normal heart but occasionally there is underlying congenital heart disease which may be subtle. An echocardiogram is invaluable in eliminating these defects (e.g. cardiomyopathy, mitral valve prolapse, Ebstein's anomaly).

TREATMENT OF ARRHYTHMIAS IN CHILDREN

MEDICAL MANAGEMENT

The introduction of radiofrequency catheter ablation and small pacemakers and ICDs has led to a reassessment of the role for long-term antiarrhythmic drug therapy in children with serious arrhythmias. In the younger child, medical management is usually appropriate for most conditions. Optimal medication depends on the age of the child, presence of coexisting disease and haemodynamic or prognostic importance of the arrhythmia. All antiarrhythmic drugs have side effects and often the long-term effects are unknown. Few antiarrhythmic medications are licensed for paediatric use. Medical management should include advice about the risks of exercise. Most children can be encouraged to participate normally but individual advice is needed and guidelines are available. [27]

Supraventricular Tachycardia (SVT)

Recommendations for the management of adults with supraventricular tachycardia have been published but these specifically exclude children.[28] SVT can usually be stopped terminated by vagal stimulation (carotid sinus massage, Valsalva manoeuvre, diving reflex in an infant) or i.v. adenosine. Both techniques act primarily by blocking conduction through the AV node. An ECG should be recorded as this may define the aetiology. Other drugs should be used with caution as they can precipitate other arrhythmias (e.g. digoxin, flecainide), and may be profoundly negatively inotropic (e.g. verapamil). In the collapsed child, cardioversion is effective and safe (1-2 Joules/kg).

Preventing recurrence of SVT using antiarrhythmic drug treatment can be more difficult. In infant SVT, it is helpful for parents to be taught to listen to the heart using a stethoscope. This allows SVT recurrence to be detected before heart failure develops. Due to pathway "maturation", 80% of infants with SVT may be symptom free by 1 year but up to 30% will have a recurrence of palpitations in later childhood. In older children, a wide variety of drugs are available. The choice is determined by side-effect profile in addition to efficacy. AV node blockers should be avoided in WPW syndrome because of the risk of precipitating antidromic AVRT. In view of these various considerations careful consideration of curative radiofrequency ablation should be made assessing the relative risks and benefits.

Ventricular Arrhythmias

The medical treatment of VT will depend on the underlying mechanism. In children with congenital heart defects, the onset of VT often reflects underlying haemodynamic problems and this should be investigated. Beta-blockers are helpful in catecholamine-induced VT and the long QT syndrome. Amiodarone is very effective in many forms of SVT and VT but use is limited by serious long-term side effects.

AV Block / Bradycardias

In the infant, bradycardias are usually secondary to respiratory compromise. In the context of a primary bradycardia, medical management is usually futile and temporary or permanent pacing is required. Most pacemakers are implanted for congenital heart block. The indications for pacemaker implantation in an asymptomatic child with AV block are:

- Average 24hr heart rate < 50/min.
- Presence of ventricular ectopy.
- Presence of Long QT interval (QTc > 440ms).

All children with postoperative heart block persisting 7-14 days should undergo pacemaker implantation because of the small risk of sudden death.[29] Some children will eventually recover conduction but long term studies are needed to establish whether a long term pacemakers will be necessary.[30] All children with congenital CAVB should undergo pacemaker implantation by age 15yrs even if asymptomatic. Long term studies have shown a 26% incidence of Stokes Adams attacks which may be fatal in up to 30%.[31 32]

ELECTROPHYSIOLOGY STUDIES AND ABLATION

Until the advent of the catheter ablation, electrophysiology studies were rarely performed in children. However, now many arrhythmias can be cured, (see Chapter 14, 16, 18).

The Paediatric Electrophysiology Society Registry reported the results of 725 ablation procedures in 1994. Success rates of 83% for both AVNRT and AVRT were reported with a complication rate of only 4.8%. Complications were higher in younger children (10%) and included death (0.5%), CAVB (1.1%) and other serious complications such as cardiac perforation/ pericardial effusion, pulmonary/systemic emboli and valve regurgitation in 1.5%. [33] Since this 1994 report, success rates have improved and complications fallen. Radiofrequency ablation procedures have been used successfully for a variety of arrhythmias (supraventricular and ventricular) in infants and children with structurally normal and abnormal hearts.[34-37] In infants, however, there is some evidence of growth of radio-frequency lesions and the procedure may be reserved for the older child and adolescent unless symptoms are life threatening and resistant to medication. [38]

PACEMAKERS AND IMPLANTABLE DEFIBRILLATORS IN CHILDREN

Modern pacemakers are smaller and much more programmable than their predecessors. Together with pacing lead miniaturisation, this has allowed endocardial pacing to be performed in young children. However, many units still implant epicardial systems in the infant to preserve venous access for future endocardial implants. Advances in lead technology (e.g. steroid elution to reduce any inflammatory reaction to the lead) have led to improvements in lead lifetime and efficiency. ICDs can be used in children at risk of life-threatening ventricular arrhythmias. [539]

THE ADULT WITH CONGENITAL HEART DISEASE

Arrhythmias are the commonest reason for emergency admission to hospital in the adult with congenital heart disease. [40 41] The arrhythmias are often atypical in presentation and a relatively slow atrial tachycardia may be mistaken for sinus rhythm. Arrhythmia management is a key factor in the medical care of this complex group of patients. Management may involve hybrid therapy with antiarrhythmic drugs, catheter ablation guided by electroanatomic mapping, antitachycardia pacing, ICD implantation and arrhythmia surgery. [15 42 43]

STRUCTURE OF PAEDIATRIC AND CONGENITAL HEART ARRHYTHMIA SERVICES

A successful congenital heart disease arrhythmia service requires:

- Close collaboration between the paediatric cardiologist, adult electrophysiologist, paediatric arrhythmia specialist and cardiac surgeon.
- Paediatric service should be situated within a paediatric cardiac unit.
- There should be adequate facilities for paediatric cardiac surgery.
- The adult congenital heart disease arrhythmia service should have close links with the paediatric cardiology service.
- Non-invasive assessment including ambulatory monitoring and implantable monitoring should be available.
- A dedicated arrhythmia medical, nursing and technical team with experience and facilities for complex pacing and electrophysiology should be available.
- Electrophysiology studies should be performed by an experienced electrophysiologist with additional expertise and experience in congenital heart disease.
- The paediatric arrhythmia specialist/cardiologist will need to work jointly with an adult electrophysiologist.
- Centres carrying out invasive electrophysiology procedures in children should comply with the Recommendations for Interventional Paediatric Cardiologists and Paediatric Cardiology Cardiac Catheterisation Laboratories[44 45]
- Electroanatomic mapping should be available for complex procedures and there should be a link with a surgeon with an interest in arrhythmia surgery in the congenital heart disease patient.
- Congenital heart arrhythmia service should have a close link to a genetics service with an interest in the genetics or cardiac arrhythmias, cardiomyopathy and the chanellopathies.

LAY SUMMARY

Children can develop many of the heart rhythm disorders seen in adults, but there are rhythm disorders that occur only in children, and there are special circumstances in children that make heart rhythm treatment more difficult in children.

Children can develop rapid heart rhythm abnormalities that persist much of the time or all the time, and when this happens, the heart may become tired and begin to fail. Recognition that the heart rhythm has caused heart failure is essential to ensure that treatment is directed at the heart rhythm in order for the heart failure to improve.

Treatments with catheter ablation, pacemakers and devices, and occasionally heart surgery,

has greatly improved the outcome of children with arrhythmias. Catheter ablation can be curative, and the relative risks and benefits should be carefully considered in the individual case in a multidisciplinary group involving paediatric cardiologists and cardiac electrophysiologists.

Management of arrhythmias in children and in children and adults with congenital deformities of the heart should only be undertaken in well-equipped and staffed centres of excellence where all the necessary specialist consultants and equipment are available, and the team of doctors treats many such patients all year round. Such concentration of skills ensures the best results.

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