A CASE OF MORBUS CAERULEUS.

By MAJOR D. G. EVANS.
Royal Army Medical Corps.

This case is reported, as such cases are not often seen in military medical practice, also the differential diagnosis of the exact pathological condition present is interesting and presents considerable difficulty.

J. B., aged 9, daughter of a quartermaster-sergeant, Royal Corps of Signals, was first seen on August 26, 1928, for measles.

Family History.—Her father, mother, four sisters, and one brother are all alive and well, and she is intermediate in age between the other children. Her mother is inclined to obesity, and states that she had a nervous shock when pregnant with this child.

Previous Health.—The patient has had bronchopneumonia twice, at the age of 9 months and again at the age of 1 year and 9 months. At the age of 3 years she had whooping-cough. On August 26, 1928, she developed measles; the attack was a mild one, but she fainted and cut her head during convalescence. At the age of 3 years and 9 months she went with her parents to Jamaica, and remained there for three and a half years, i.e., until about two years ago. Her mother states that the climate appeared to agree with her.

History of the Present Condition.—The heart condition from which she suffers was first recognized when she was 10 months old by a medical practitioner at Farnborough. She has always been more or less short of breath, especially on exertion, such as hurrying or running up hill, and has not played out-door games for this reason. In cold weather she also becomes blue about the lips. She gets a funny feeling occasionally at night with a sensation at the lower end of the sternum; the parents say that this is usually relieved by alcohol.

Present Condition.—She is rather short of breath on exertion, but walks uphill to school daily, and has even been seen to run downhill.

General Appearance.—The lips and nails show a slight degree of cyanosis. There is a little clubbing of the fingers and toes, but it is not very noticeable.

The Circulatory System.—(1) The pulse-rate is 85, the beat is regular and of rather low tension, both radial arteries beat synchronously. (2) The
Clinical and other Notes

heart: The apex-beat is in the fifth left interspace, four and a half inches to the left of the midsternal line. There is a distinct systolic thrill felt in the region of the second left interspace. The area of deep cardiac dullness extends downwards from the third rib, the right border is two-thirds of an inch to the right of the right border of the sternum, and the left border is through the line of the apex beat. Heart sounds: There is a murmur, systolic in time, and of an extremely harsh and blowing quality, heard best over the pulmonary area (second left interspace). This murmur is transmitted obliquely upwards to the left clavicle, but is also heard all over the heart region in front, as well as below the angle of the left scapula behind. The murmur entirely replaces the first sound; the second sound is heard, but is weak.

X-ray.—The heart shadow is greatly enlarged to the left side, almost
Clinical and other Notes

obliterating half the left lung; the right border is seen nearly an inch to the right of the sternum.

Blood.—Total red cells, 4,620,000 per cubic millimetre; percentage of haemoglobin, 85; colour index, 0.9.

Alimentary System.—No enlargement of the liver or spleen was detected. Other systems normal.

Differential Diagnosis.—A systolic murmur of maximum intensity at the pulmonary area may be due to: (1) congenital pulmonary stenosis with or without defects of the interauricular septum (patent foramen ovale), or the interventricular septum; (2) a functional bruit (as in severe anaemia); (3) acquired pulmonary stenosis (due to the endocarditis of rheumatic fever, etc.).

(1) Congenital Pulmonary Stenosis.—In favour of this is the character of the murmur and the discovery of the lesion at the age of 10 months. It is to be remembered that pulmonary stenosis is the most frequent of congenital lesions. This diagnosis is favoured by the presence of the thrill, the considerable enlargement of the right side of the heart, together with cyanosis of varying degree and the clubbing of the fingers and toes—although this clubbing is at present only slight in degree.

(2) A Functional Bruit.—Against this is the history, the quality of the bruit, and the presence of a distinct thrill, the large size of the heart, also the bruit does not alter with the position of the patient (a functional bruit is louder in the recumbent than in the erect posture).

(3) Acquired Pulmonary Stenosis.—Against this diagnosis is the great rarity of this condition and the history of the early discovery of this lesion, and the fact that endocarditis is stated to be rare in infants. The absence of a history of rheumatic fever and the presence of cyanosis and clubbing are also against it.

(4) Patent Ductus Arteriosus (Ductus Botalli) may also be mentioned, but in this condition by itself the murmur is prolonged into diastole, and does not give rise to clubbed fingers—cyanosis is also usually absent. In such cases the symptoms may be very slight, and are not incompatible with a healthy life, and the condition may only be accidentally found out. A long rumbling bruit commencing during ventricular systole, and passing on into the diastole of the ventricles, is considered to be pathognomonic of this defect, as it is impossible for a bruit extending from systole into diastole to be produced within the heart. When this condition occurs with pulmonary orifice defect, it is found that the defect is nearly always complete atresia (Greene).

(5) Associated Septal Defects.—Congenital pulmonary stenosis is nearly always (90 per cent) associated with patent interventricular septum, and it is also often associated with a patent foramen ovale. Trivial foraminal defects may be present, and are common. The blood then partly flows from the right ventricle into the pulmonary artery, but owing to the narrowing of the pulmonary artery, some of the blood has an alternative course, and
may pass from the right auricle or ventricle direct to the left auricle or ventricle respectively through the patent septum and thus escape oxygenation. An open foramen ovale may yield a presystolic murmur, or no signs may be present at all during life. With an associated ventricular septum defect, both the thrill and the murmur may be more intense over the third and fourth left interspace and lower sternum.

**Diagnosis.**—It is considered that congenital pulmonary stenosis is undoubtedly present, and probably a patent interventricular septum also—though this cannot be stated with certainty. More is often found after death in these cases than was ever suspected during life. Such cases were formerly called blue babies or congenital cyanosis, and are also called cases of morbus caeruleus.

**Etiology and Pathology.**—This condition is said to have its inception usually during the second month of foetal life. It is considered that the lesion usually results from arrest of development at the conus arteriosus on the heart side of the valves, or beyond the valves in the pulmonary artery itself, where the trunk is in close relation to the aorta. In some few cases it results from foetal endocarditis. It is almost invariably associated with a patent ventricular septum.

**Prognosis.**—She is unlikely to reach the end of the second decade, and few survive the first seven years. If there is no septal defect, she has a good chance of reaching the third decade of life. With ventricular septum and foramen ovale alike patent she would be expected to die during the first two decades.

**Treatment.**

(1) **General.**—The patient may be allowed to continue to go to school, but she should always take her own time and go her own pace at any walking or other bodily exertion. She should avoid competitive outdoor games, and should realize that she cannot do the same as other children. In due course should she live to be a marriageable age, she should be warned against marriage, owing to the dangers of pregnancy and lactation. If she were to become pregnant, it is considered that abortion should be induced early—although Sir James Mackenzie mentions a case who successfully underwent a confinement at full term. She should be kept under regular medical observation in order to detect, and as far as possible to prevent, the onset of cardiac failure. The patient should be protected against respiratory and other intercurrent diseases.

(2) **Climatic Treatment.**—It is considered that moderate cold is stimulating to the heart and great heat is depressing; accordingly a fairly dry, cold climate is best, and a damp climate should be avoided if possible. The patient should not return to the tropics, and should avoid all hill stations and hilly places if this can be done. Dover is rather hilly, but the patient appears to be doing well here.

(3) **Drugs.**—No drugs are required at present, for occasional discomfort at night alcohol appears to be useful. If cyanosis should become severe, it is considered that warm inhalations of oxygen gas should be given.