The Budd-Chiari syndrome

Oclusion of the hepatic veins is a rare condition usually caused by tumour or thrombus arising either locally or by extension from the inferior vena cava. The role of congenital lesions is disputed. The aetiology remains unknown in over two-thirds of patients.

Intrahepatic venous occlusion can follow the ingestion of certain alkaloids, particularly senecio (veno-occlusive disease). These are taken in herbal medicines, particularly in the West Indies, India and the Middle East. Plant toxins are probably important causes of hepatic injury.

Phlebitis of the centrilobular hepatic veins can also follow radiation applied to the liver and also the use of cytotoxic drugs such as urethane.

The condition should be suspected if a patient with a tendency to thrombosis or with malignant disease in or near the liver develops tender hepatomegaly with gross ascites.

Special radiological investigations (Clain et al., 1967) are essential to confirm the diagnosis. Hepatic venography may fail or show narrow occluded hepatic veins. Adjacent veins show a tortuous, lace-like spider-web pattern. These probably represent abnormal venous collaterals. The catheter cannot be advanced the usual distance along the hepatic vein and wedging occurs 2–12 cm from the diaphragm. The features of a normal wedged venogram are absent. Inferior vena-cavography establishes the patency of the inferior vena cava. The hepatic segment of the vein may show side-to-side narrowing due to a large liver. Splenic venography may show a collateral circulation. Selective coeliac arteriography shows a small hepatic artery with branches of fine calibre.

Hepatic scintiscans are abnormal with poor uptake of isotope into the areas drained by the occluded hepatic vein. The caudate lobe may show good uptake.

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References


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The management of the coronary crisis

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Until recently the death rate of patients admitted to hospital following an acute myocardial infarction was as high as 30%. The cause of death is not always clearly understood and in order to increase our knowledge and reduce the appalling mortality in this condition Sir John McMichael in 1962 suggested making a detailed study of the haemodynamic and other changes taking place in such patients. Little work had previously been done in this direction as it had been considered that the patients were too ill to study in detail; in many cases treatment
was given without a clear understanding of the circulatory and other changes following a myocardial infarction. With Sir John's encouragement and in association with the Medical Research Council a side-ward in Hammersmith Hospital has been equipped with monitoring apparatus so that the haemodynamic, respiratory, renal, metabolic and other changes can be followed at the side of the bed without disturbing the patient. Data collected in this ward are relayed to a central recording room and continuously recorded over the 24 hr on magnetic tape which may be subsequently analysed. As a result it has been possible to put the management of the patient following an acute myocardial infarction on a more rational basis. This, together with the introduction of defibrillation by electric shock, electrical pacing of the heart and the concept of 'intensive care' has considerably reduced the mortality in patients admitted to hospital.

The cause of death following myocardial infarction may be the development of an arrhythmia, failure of the myocardium as a pump due to extensive involvement by the infarction, biochemical changes, secondary respiratory or renal impairment, embolism or the misuse of drugs.

The management of the arrhythmias

Continuous recording of the electrocardiogram on magnetic tape has shown that some form of arrhythmia occurs in 80% of all patients at some time in the first few days following a myocardial infarction. The arrhythmias are often transient and may last from a few seconds to hours or days. Their unpredictable behaviour often with spontaneous recovery makes assessment of the value of drugs in their treatment difficult. In many cases where the arrhythmia does not signify the risk of the development of more serious complications no specific treatment is indicated. In others the right treatment may be life-saving. Arrhythmias may be classed under the headings of extrasystoles, supraventricular and ventricular bradycardia, supraventricular and ventricular tachycardias, disturbances in atrio-ventricular conduction and asystole.

Extrasystoles

Atrial and ventricular extrasystoles are common following myocardial infarction and if occurring infrequently may be ignored. If, however, they occur more frequently, early in the cardiac cycle with interruption of the T wave, in salvos of two or more, show a multiform configuration, or occur in a frequency greater than five per minute, treatment should be given as these may herald the onset of more serious arrhythmias such as ventricular tachycardia or ventricular fibrillation. A single injection of 25 ml of 2% Xylocaine is given in the first place. If the arrhythmia is not suppressed a further 50 mg is given. If this treatment suppresses the ectopic beats a continuous intravenous drip of from 1 to 2 mg of Xylocaine diluted in glucose and water a minute is given. Usually the drip has to be maintained for 24-48 hr when the ectopic beats will have disappeared spontaneously (Fig. 1).

In the rare case where Xylocaine is ineffective, 50 mg of procaine amide may be given intravenously every 2 or 3 min until the arrhythmia is abolished, followed by an oral maintenance dose of 1-2 g daily for 2 or 3 days.

Nodal rhythm and supraventricular bradycardia

Atrio-ventricular or nodal rhythm may develop following myocardial infarction with consequent loss of atrial transport function. In these rhythms the normal sequence of atrial and ventricular contractions is distributed to varying degrees, depending on the altered relationship of the P and QRS waves in the electrocardio-
Supraventricular bradycardia, or slowing of the heart in sinus rhythm occurs at some stage in 20% of all patients following an acute myocardial infarction and may lead to a profound shock syndrome. This form of bradycardia may occur spontaneously and at times appears to be potentiated by the administration of morphine or other analgesic drugs. When the bradycardia is moderate and transient it is usually of little clinical significance. More serious consequences follow if it is profound and particularly if ventricular function is poor with low stroke volume. Symptoms may be limited to a weak sensation and nausea while lying flat but with dizziness, pallor and sweating after sitting up or standing. The general appearance resembles that of a common 'vasovagal' faint. If the changes progress, mental confusion may be succeeded by loss of consciousness associated with the clinical picture of profound circulatory shock. Cardiac arrest may follow.

Treatment is indicated in most patients at an early stage in view of the potential haemodynamic deterioration and also because of the increased incidence of serious arrhythmias occurring in patients in whom bradycardia is allowed to persist. Mild cases may be managed conservatively by maintaining the patient flat and raising the foot of the bed. If the heart rate does not increase and the blood pressure remains low, atropine sulphate should be injected intravenously. In the first place 0·3 mg is given and the pulse rate recorded for 5 min, if it has not reached 80–90 beats/min a further 0·3 mg is given with similar increments, if necessary up to a total of 2 mg (Fig. 2). Subsequent doses may be necessary 2 or 3-hourly until the heart rate is maintained at normal levels. It must be emphasized that the drug should be given intravenously as absorption from subcutaneous or intramuscular injection is too slow and unpredictable under conditions of circulatory failure.

Heart-block

Incomplete or complete heart-block is a not infrequent complication of myocardial infarction and where complete may be serious. Measurement of the stroke output of the heart under
these conditions has shown that the damaged ventricle is not able to compensate for the slow heart-rate by increasing the amount of blood expelled at each beat and a falling cardiac output rapidly leads to circulatory impairment.

It is not necessary to treat first-degree heart-block. For second degree heart-block the drug of choice is isoproterenol hydrochloride. A 1 mg ampoule diluted in 500 ml of dextrose in water is administered intravenously at a rate sufficient to raise the ventricular rate above 60 beats a minute. Care must be taken not to give too much isoproterenol which may produce ectopic beats and herald the onset of ventricular fibrillation. In complete heart block the heart must be paced electrically by passing an electrode catheter into the right ventricle via an arm or neck vein. Steroids may be given although it is not clear how effective they are.

**Ventricular tachycardia**

Ventricular tachycardia is a frequent, and one of the most serious, of the arrhythmias, and if persistent demands immediate treatment. Often, however, it presents as brief paroxysms which are self-limiting. Treatment is the same as for ventricular ectopic beats and calls for the administration of a 50 mg injection of Xylocaine followed by a 100 mg injection if the first does not succeed in establishing sinus rhythm. If the ventricular tachycardia persists, electrical cardioversion starting at 50 msec must be employed at once.

**Ventricular fibrillation**

The recent introduction of external cardiac massage and electrical defibrillation has been responsible to leading, in part, to the establishment of intensive care units where the patient may be constantly watched and the development of ventricular fibrillation immediately treated. In the occasional case the institution of external cardiac compression may revert the patient to sinus rhythm but in nearly all it is necessary to use an electrical defibrillator starting at 100 msec and increasing the shock by 100 msec up to 400 in the occasional case. Patients who have cardiac arrest develop a profound acidosis which must be corrected by intravenous sodium bicarbonate. This may be controlled by estimating the pH of the blood.

**The management of a failing circulation unassociated with an arrhythmia**

In some cases a failing circulation associated with a falling blood pressure, the development of pulmonary oedema, cold peripheries, oliguria and mental impairment or coma may occur in the presence of sinus rhythm. This is almost invariably the result of failure of the damaged ventricle to maintain its stroke output and is usually accompanied by a tachycardia. Recent work has shown myocardial failure to be accompanied by a very great increase of catecholamine, both adrenaline and noradrenaline, excretion. The administration of pressor agents which, although they may raise the blood pressure, will further impair cardiac function and regional organ perfusion, is contraindicated. Treatment should be directed to improve the performance of the damaged left ventricle although in some cases this may be so extensive that the residual muscle is insufficient to maintain an adequate cardiac output and treatment will be of no avail. The administration of oxygen together with a diuretic will improve the oxygenation of the damaged muscle. Digitalis may be given but recent work has shown that this may have little

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**Fig. 4.** With the development of complete heart-block there is often a serious fall in the cardiac output and blood pressure.

**Atrial fibrillation**

Atrial fibrillation, lasting from a few seconds to hours, or infrequently, days, can often be seen on a continuous recording of the patient's electrocardiogram. If there is evidence of circulatory embarrassment associated with the rapid heart rate, digitalis should be given to slow the rate. This may be administered in an intravenous form and continued by mouth.

**Supraventricular tachycardia**

Supraventricular tachycardia may occur in short bursts in the acute stage of myocardial infarction and most often spontaneously reverts. In the event of its persisting with a deterioration in the patient's condition, the method of treatment if available is electrical cardioversion followed by a maintenance dose of procaine amide or propranolol.
or no effect on cardiac performance in the acute stage of myocardial infarction. The reflex changes occurring in the peripheral and venous circulation are not yet clear in these patients and much more research is needed under full haemodynamic and other control to evaluate the value of drugs, the administration of intravenous fluids and other recommended procedures in these seriously ill patients.

Other factors in the management of the patient following acute myocardial infarction

Posture

In many cases it probably makes little difference whether the patient is nursed flat or in the sitting position. Patients with sinus bradycardia due to the vaso-vagal syndrome should be nursed flat with the foot of the bed raised. Conversely those with severe left ventricular failure and pulmonary oedema are better sitting up. In all cases a commode is preferable to a bed-pan for movement of the bowels.

Oxygen

Recent work has shown that the oxygen tension in the blood is lowered in the majority of patients following an acute myocardial infarction. Respiratory function tests have shown this to be due both to diffusion impairment in the alveoli possibly due to incipient or frank pulmonary oedema and to uneven ventilation/perfusion due to patchy atelectasis. In most cases the blood oxygen tension can be raised to normal levels by the administration of 30% oxygen by means of a simple mask. Haemodynamic studies of the effect of oxygen have demonstrated a rise of arterial pressure associated with a fall in cardiac output.

In view of these findings the continuous administration of oxygen throughout the acute stage of the illness may be beneficial and should always be given in cases where the circulation is grossly impaired.

Blood electrolytes

Although significant changes in blood electrolytes are not the rule following myocardial infarction it is important to watch and control the level of blood potassium especially in cases of renal impairment.

The use of other drugs

The control of pain in the early and acute stages of the disease presents a special problem in itself. In most cases morphine sulphate is the drug of choice but it must be remembered that this may potentiate a vaso-vagal attack in those patients sensitive to it, accompanied by nausea, vomiting, bradycardia and hypotension which in turn may be controlled by atropine. The excessive use of morphine may lead to respiratory depression with impairment of respiratory function. It may well be that heroin with its relative freedom from side-effects may in some cases be a better analgesic drug for these patients. Where the pain is not so severe pethidine may be used. The question as to whether anticoagulants should be used is still an open one and has not been conclusively settled either way. There certainly was no striking decrease in mortality following their introduction. In the intensive care unit at Hammersmith Hospital anticoagulants are only given where there is clear evidence of venous thrombosis in the leg veins.

Intensive care units in the treatment of acute myocardial infarction

The information gained by the intensive monitoring of the electrocardiographic, haemodynamic and other changes taking place in the patient following a myocardial infarction with the resultant improvements in our understanding and treatment of the condition together with the constant care of patients in intensive care units appears to have reduced the mortality by about a third in the last 4 years. It is probably still too early to say how such units will develop in relation to medicine as a whole in view of the rapidly changing methods of management of such complications as the arrhythmias. A good beginning has been made but much further research has to be done to improve our knowledge of this disease and the correct application of treatment for its complications.

Selected references


