Editorial

A Program for Stokes-Adams Disease and Cardiac Arrest

In the last 10 years interest in Stokes-Adams disease has been renewed and many new approaches have been developed. Formerly, it was considered a rare disease of the aged for which the prognosis was poor and the treatment was unsatisfactory. Now, it seems more common and is seen not infrequently in young people. Furthermore, new types of treatment provide solutions for many of the problems of Stokes-Adams disease and even offer the possibility of complete prevention of seizures. These new developments include external electric cardiac stimulation and countershock, cardiac monitoring, intravenous administration of sympathomimetic amines, mouth-to-mouth breathing, hypothermia, short-term and long-term direct electric stimulation and external cardiac compression.

Elsewhere in this issue is described an effective pharmacologic approach to one of the remaining problems of Stokes-Adams disease, the prevention of recurrent attacks due to ventricular tachycardia and fibrillation. Epinephrine and isoproterenol, despite their well-known excitatory effects on ventricular rhythmicity, were found to prevent ventricular irritability in Stokes-Adams disease.

Stokes-Adams disease poses three major problems that require different therapeutic approaches (table 1): the prompt restoration of circulation in the emergency of cardiac arrest; the restoration of intrinsic cardiac rhythm; and the prevention of recurrent episodes. A program for the management of these three problems is applicable to Stokes-Adams disease and indeed to cardiac arrest of any origin.

Restoration of Circulation

Circulation and respiratory exchange must be restored within the stringent time limit of 3 or 4 minutes before the brain and heart have suffered irreversible damage. This is the cardinal problem not only in Stokes-Adams attacks but in cardiac arrest of all kinds. It is therefore necessary that a program be prearranged and well rehearsed and that the arrest be recognized promptly. Continuous cardiac monitoring with a combined monitor and external electric pacemaker permits immediate recognition and treatment, and has been particularly valuable in Stokes-Adams disease and during anesthesia.

The sequence of procedures used to restore circulation is based on their relative safety and speed of applicability (table 1). If a monitor-pacemaker and external countershock defibrillator are immediately at hand, as is often the case in Stokes-Adams disease and in the operating room, resuscitation from ventricular standstill or fibrillation is ordinarily effected promptly. If this equipment is not immediately available or is not effective, a few precordial blows and then external car-
Table 1

The Three Problems of Stokes-Adams Disease and Cardiac Arrest, Program for Their Management

1. Restoration of circulation
   - Precordial blow
   - External electric stimulation or countershock
   - Thoracotomy and massage
     - If other methods fail
     - If adequate help is available
     - If prognosis is favorable: No irreversible cerebral damage
     - General condition good
     - Artificial respiration

2. Restoration of intrinsic cardiac rhythm
   - Electrocardiograph or monitor
   - Electric stimulation or countershock
   - Drugs: epinephrine, isoproterenol, proeaine amide, calcium salts, norepinephrine, sodium bicarbonate

3. Prevention of recurrent episodes
   - Intravenous epinephrine, isoproterenol, atropine
   - Oral epinephrine and isoproterenol
   - Internal electric pacemaker

diac compression should be applied. The application of external electric currents for stimulation or countershock or of several blows to the precordium carries very little risk, whereas external cardiac compression may produce significant injuries. Cardiac puncture with or without the intracardiac injection of epinephrine, though hazardous also, may be tried next if the preceding measures fail.

Thoracotomy and direct manual compression of the heart remain as the procedures of last resort for the emergency restoration of circulation. With the frequent success of external cardiac compression thoracotomy and direct massage are required much less frequently. Failure of external compression, however, does not necessarily mean that circulation cannot be restored by other means; thoracotomy and massage have at times been successful in this desperate situation. Thoracotomy should rarely be necessary in Stokes-Adams disease but may be considered at times in other types of cardiac arrest. This heroic approach should be undertaken only if the other procedures have failed or cannot be applied promptly, if adequate help with the thoracotomy and with respiration is quickly available, and if the time limit for cerebral recovery has not been exceeded and the patient’s outlook is otherwise good.

If spontaneous respiration ceases, artificial respiration by any appropriate means becomes a vital part of the resuscitative effort. The two major requirements for adequate respiratory exchange are a clear airway and intermittent positive pressure. The latter can be provided promptly by mouth-to-mouth breathing.

Restoration of Intrinsic Cardiac Rhythm

The emergency restoration of circulation is often attained or followed promptly by return of effective intrinsic ventricular activity. At times, however, in all types of cardiac arrest, effective intrinsic cardiac rhythm may not return. Although the patient is being kept alive by artificial means, such as external electric or mechanical stimulation and external or direct cardiac compression, cardiac resuscitation is not complete until this second problem has been met successfully.

Since circulation to the heart and brain is being maintained, there is adequate time to determine the mechanism of the cardiac arrest with an electrocardiogram and to treat it appropriately. Persistent ventricular standstill may require prolonged external electric stimulation and the intravenous administration of dilute solutions of epinephrine or isoproterenol for the arousal, acceleration, and maintenance of an intrinsic ventricular pacemaker. The choice between epinephrine and isoproterenol, which have similar effects on ventricular rhythmicity, is usually made on the basis of their effects on the blood pressure. Ordinarily, epinephrine produces a striking rise in pressure, whereas isoproterenol has little or no effect. The drugs are given in dilutions of 4 mg. per liter at initial rates of 15 to 30 drops (4 to 8 micrograms) per minute with stepwise increases every few minutes until the desired effects or toxicity (excessive acceleration, ectopic activity, hypertension) appears. Persistent ventricular fibrillation may require external or direct countershock and the intravenous or intracardiac administration of drugs like epinephrine (0.2 to 0.5 ml).
of 1:1000 solution), procaineamide (50 to 200 mg. at intervals of 1 to several minutes), and calcium salts (4 to 6 ml. of 10 per cent calcium gluconate) to terminate fibrillation, to prevent recurrences, and to improve myocardial contractility.

In addition, blood pressure, blood pH, and respiration should be maintained by appropriate measures such as intravenous norepinephrine (continuous infusion of 4 to 8 mg. per liter) and sodium bicarbonate (3.75 Gm. in 50 ml. intravenously and repeated as necessary), and intermittent positive-pressure breathing of oxygen by bag and endotracheal tube. Finally, once cardiac resuscitation has been completed, hypothermia and agents like urea (1.0 to 1.5 Gm. per Kg. in 30 per cent solution intravenously in 1 to 2 hours) to reduce cerebral edema should be used if consciousness does not return promptly.

Prevention of Recurrent Episodes
Management of the two preceding problems completes the resuscitation from cardiac arrest but the threat of recurrent episodes remains. After arrest of any kind the cardiac rhythm should be monitored continuously, and emergency readiness to manage recurrence must be maintained for at least 72 hours. In Stokes-Adams disease, in which seizures may recur unpredictably for the rest of the patient’s life, special preventive measures are required.

Drugs may prevent Stokes-Adams attacks by stabilizing or improving atrioventricular conduction, by accelerating and maintaining idioventricular pacemakers, and by reducing ventricular irritability. Administered intravenously in dilute solutions, epinephrine and isoproterenol usually prevent recurrent attacks for periods of days. As reported in this issue, this technic is effective in preventing recurrent ventricular tachycardia and fibrillation as well as recurrent standstill. Recurrent seizures may also be prevented by electric stimulation at adequate rates. This can be accomplished for the short term either externally or with a catheter electrode in the right ventricle. The latter approach, though painless, carries more hazards (ventricular fibrillation, cardiac perforation, ineffective stimulation, thrombosis, sepsis) than does treatment with drugs. In this connection, it should be re-emphasized that depressing drugs such as quinidine, procaine amide, and potassium salts are contraindicated in patients with complete heart block; if given to Stokes-Adams patients with ventricular ectopic beats, tachycardia, or fibrillation, they may aggravate rather than prevent seizures. Finally, in an occasional patient, atropine (1.0 to 2.0 mg.) given intravenously may stabilize atrioventricular conduction or accelerate and maintain a ventricular pacemaker.

In contrast to the effectiveness of intravenously administered sympathomimetic amines for the short-term prevention of seizures, drug administration orally for the long term is less reliable. Ephedrine (25 to 200 mg. every 4 hours by mouth) and isoproterenol (5 to 40 mg. every 2 to 4 hours sublingually) are at times most useful. Corticosteroids and chlorothiazide are often recommended for this purpose, but they appear to us to be of little value.

For assured prevention of attacks, whether due to ventricular standstill or fibrillation, the unreliable intrinsic ventricular pacemaker may be replaced by an electric one to drive the ventricle continuously and indefinitely. Sufficient experience has now accumulated to indicate the practicability of this approach in eliminating the threat of recurrent Stokes-Adams attacks. In patients with heart block internal electric pacemakers may now also be recommended for the treatment of congestive heart failure due to slow ventricular rates.

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References

William Harvey

Love of truth, reverence and charity, with some tincture of imagination and humour, these were the chief features of Harvey's personality; but to complete the picture we must add moral courage—which Michael Foster said is an essential part of the scientific character—patience and reflectiveness. It must have required much moral courage to attack the Galenic stronghold, and it is no wonder that Zachary Wood apostrophised him as “Truly a bold man, indeed, O disturber of the Quiet of Physicians! O Seditious Citizen of the Physical Commonwealth!” His patience was shown not only in the carrying out of his investigations, but in his reluctance to make them public. He always bided his time. He was like Darwin, who waited 29 years for the results of a single experiment, and all of whose work on evolution was published after he was 50. In his slowness to publish he resembled other great men of his time, and the next century, Galileo, Newton, Bacon, Cavendish and Gauss. How different from us today with our “preliminary notes” and disputes as to precedence in discovery! Lastly, he was a reflective philosopher. Like Hunter his delight was to think. We are told that he would withdraw himself to the leads of his house in town, or to caves in his garden in the country, in order to indulge in contemplation. Surely in this also he has a lesson for our unreflective time.—Sir Robert Hutchison (Harveian Oration, 1931). The Quiet Art: A Doctor's Anthology. Compiled by Dr. Robert Coope. Edinburgh & London, E. & S. Livingstone Ltd., 1952, p. 54.

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