Choreic syndrome after cardiac surgery

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SUMMARY Five children had an acute choreic syndrome that started three to seven days after hypothermic cardiopulmonary bypass surgery. Improvement after an interval was seen in all, with complete resolution in two. In addition to the chorea, all children suffered complete supranuclear ophthalmoplegia, which has also shown subsequent improvement. Bulbar damage caused considerable speech problems. There was no cognitive impairment. Several drugs were tried without effect. Investigations of the cause, including histological examination, have been unrevealing.

The onset of acute chorea is both dramatic and profoundly disturbing; it can arise in various settings. The clinical signs of carbon monoxide poisoning, Wilson’s disease, Leigh’s disease, local tumours, inflammatory processes, demyelinating or ischaemic lesions are not difficult to understand, because they originate in the basal ganglia. In classic post streptococcal Sydenham’s chorea the explanation may lie in the fact that the cytoplasm of the cell bodies in the caudate and subthalamic nuclei seem to be antigenically similar to the streptococcus.1 Chorea precipitated by pregnancy or the contraceptive pill may be a result of the effect of oestrogens on dopaminergic activity of basal ganglia.

Chorea after hypothermic cardiac bypass has occasionally been described,2–7 but only in childhood. We report five additional children with a strikingly stereotyped presentation and course, having in common intriguing additional clinical features. We hope that by drawing attention to this distressing condition we will encourage others to report similar cases and thereby stimulate attempts to identify its cause.

Patients and methods

Five cases presented at the Brompton Hospital during an 18 month period from July 1985. Cardiological data were extracted from the hospital records by careful scrutiny of the anaesthetic and operative records. Cases 1–4 were under the neurological care of ROR, and details of the fifth child were taken from the case notes of the Hospital for Sick Children, Great Ormond Street.

Results

Ages at the time of cardiopulmonary bypass ranged from 4 months to 6 years. Except for the one child with Down’s syndrome (case 5) all children had developed normally before operation. Cardiac defects in the individual cases are shown in table 1 together with the total time spent on cardiopulmonary bypass, the time of circulatory arrest, and the minimum core temperature. The operation was without untoward incident in all cases, as was the immediate postoperative recovery. The onset of chorea occurred after a latent period of clear normality varying between three to seven days after operation. One child (case 1) was sufficiently recovered to be discharged home before the onset of the chorea, and another (case 4) was noted to be enunciating clearly around the endotracheal tube before extubation at five days. The onset of chorea was comparatively abrupt, but its frequency and amplitude increased over the first three to five days. It then remained continuous while the child was awake, being sufficiently disturbing to preclude sleep, during which the movements disappeared. Trials of several drugs including haloperidol, tetra-benzazine, sodium valproate, benzhexol, and carbamazepine were ineffective unless given in sufficiently large doses to induce sleep. Improvement in the chorea began at periods varying from two days to eight weeks (table 2). The chorea had resolved completely after two weeks and six months in cases 2 and 4, respectively, but was residually present but continuing to improve in the other two survivors (cases 1 and 3) by nine and 19 months.
respectively. Both regained the ability to walk and feed themselves, albeit unstably. The child with Down’s syndrome died unexpectedly at home five months after operation, at a time when the chorea was still present although diminishing.

The onset of chorea was accompanied by a generalised loss of tone. With the exception again of the youngest child this persisted (although diminishing) in the other three survivors. Deep tendon reflexes in all were initially difficult to obtain. In the youngest child there was a visible loss of anal tone, which recovered after 72 hours. The oldest child became incontinent for three months, and case 3 still had urgency of micturition nine months later.

At the same time as the onset of chorea all children suffered a complete supranuclear ophthalmoplegia. Doll’s eye reflexes remained full but voluntary directed gaze was lost. With the exception of the youngest child (in whom return of voluntary eye movements was the first sign of recovery) improvement of voluntary eye movements began some time after the onset of recovery of chorea (table 2). During the recovery period there was a decreasing latent period before the onset of voluntary eye movement, which was then interrupted by coarse saccades. Smooth tracking was achieved in only two patients (cases 2 and 4). Vertical eye movements seemed to recover at the same rate as horizontal eye movements.

All children experienced difficulty with feeding; the onset of this was abrupt in all but the eldest, in whom it evolved over a three week period. In this child the tongue became contracted and comparatively immobile. In the remainder our impression was that the bulbar chorea precluded effective coordination of sucking and swallowing. Improvement in feeding dated from the improvement in chorea elsewhere.

Understanding of speech and situation remained intact throughout in all. There was no evidence of mental impairment in any case. Production of speech was, however, severely impaired in two (cases 3 and 4) despite complete resolution of the chorea in case 4, and remained mildly impaired in
case 1, 19 months after operation. The youngest child who has completely recovered is still only 13 months old.

Laboratory tests at the time of the acute onset of illness in all included routine haematological investigations, estimation of the concentrations of electrolytes (including calcium and magnesium) and blood glucose, liver function tests, and clotting studies. Results were normal in all cases. Similarly, examination of the cerebrospinal fluid (in four) and computed tomography of the brain (in three) were also normal. Additional investigations including estimation of neurotransmitter metabolites and concentrations of folic acid in the cerebrospinal fluid, ammonia in the blood, erythrocyte sedimentation rate, and aspartate transaminase activity were all normal as were electroencephalography and measurement of auditory and visually evoked potentials.

Neuropathological examination of the brain of the child with Down’s syndrome showed no features in addition to those of Down’s syndrome. In particular there was no abnormality of the basal ganglia.

Discussion

Although neurological complications after cardiac surgery are often discussed together they are clearly a disparate collection of diseases with different causes and varying outcomes. For many, the cause is readily apparent being either embolic, hypoxic or ischaemic, or haemorrhagic, and the lesion is usually visible on computed tomography. The clinical picture is defined by the vascular territory or parts of the brain involved. Thus with the exception of severe and diffuse hypoxic or ischaemic damage, the distribution of neurological signs varies from case to case.

Chorea following cardiopulmonary bypass does not come into any of these classifications. Although transient, comparatively mild unwanted movements are not uncommon after cardiac surgery, the full blown syndrome as described here is rarely reported. Because the clinical picture is so striking it cannot go unrecognised and is therefore unlikely to be under-reported. The first description we can find was by Bjork and Hultquist in 1960. They reported the neurological effects of deep hypothermia in eight children and three adults. Only two children in their series seemed to have survived operation.

Three of their children who died bore some resemblance to our cases with extrapyramidal signs appearing after the first few days. Clinical details, however, are sparse. Since then a further 12 cases have been reported, the most detailed clinical description being that of Brundberg et al in 1974, who recognised four such children in a series of 22 infants aged less than 24 months.

The clinical picture of these 17 and our five cases is rather stereotyped. No case to our knowledge has been reported outside early childhood, and the onset is from two to seven days after operation. Though it is possible that in some of these cases the onset may have been suppressed by necessarily heavy sedation in the immediate postoperative period, in most there was a clear period of normality beforehand. The severity of the chorea progressed over one to three weeks with the exception of our case 2 (the youngest child so far reported) who began to recover after 24 hours, and affected the face, bulbar muscles, and all four limbs. Where clinical details are sufficient to allow a conclusion, the tone seemed to be poor both in trunk and limbs, with reflexes either normal or suppressed. Drawing only from the previous clinical series some recovery seems to have taken place in all, with three previous cases recovering completely. Follow up periods vary, however, and insufficient information is currently available on which to base a firm prognosis.

One of the most unusual features of our cases was the oculomotor apraxia. Directed gaze in these children was only possible initially by movement of the whole head. Subsequently there was a noticeable lag before the onset of conjugate deviation with scanning, interrupted by coarse saccades; although consistent in our cases, this has not previously been noted. Because, however, note was made of ‘random’ or ‘irregular shifting’ eye movements it may have been present in other series but misinterpreted.

An apractic element may limit other functions accounting particularly for the speech delay. This might be difficult to distinguish in the younger children in the presence of unwanted movements. Our case 4 who was 6 years old at the time of surgery was able to mimic a wide repertoire of mouth and tongue movements at a time when he was virtually anarthric. During this period he was able to copy a written sentence accurately, but unable to organise the same sequence of words on a blank page. Dressing apraxia was not a feature nor did use of eating utensils or scissors provide any difficulty in addition to that expected from his chorea.

It is possible to think of a wide variety of possible pathogenetic mechanisms, including undetected hypoxia, or reduced perfusion from undetected hypocarbia or hyperviscosity. Immunological explanations entailing release of cardiac antigen and breakdown of blood brain barrier are difficult to sustain in the face of the short latent period before
the onset of the movement disorder. Idiosyncratic reactions to drugs (including anaesthetic agents) are similarly untenable. Although a variety of drugs and pressor agents were used no one agent was common to all. All children had undergone previous procedures necessitating the use of anaesthetics, and one child had had a previous hypothermic bypass procedure (case 4).

Because computed tomography in the three of our cases in which it was carried out and in both Diebler and Dulac’s two cases was normal, structural damage if it occurs must be extremely focal and discrete; we failed, however, to identify any histological changes in our one case that came to necropsy.

The latent period is equally difficult to account for. The timing before onset is similar to that of delayed onset hypoxic encephalopathy, and the pathogenesis is similarly obscure. It is conceivable that sufficient cellular damage occurs to interrupt neurotransmitter synthesis or reuptake, the onset of symptoms coinciding with depletion of intracellular stores. Chorea has been attributed to excessive dopaminergic activity. This excess activity may, however, only be so in comparison with that of other neurotransmitters such as acetyl choline. The suggestion of, for example, impermanent depletion of basal ganglia and frontoparietal acetyl choline is one possible explanation consistent with normal concentrations of 5-hydroxy indole acetic acid, 5-methoxy 4-hydroxy phenyl glycol, and homovanillic acid in cerebrospinal fluid, the ineffectiveness of therapeutic trials to date, and the oculomotor apraxia.

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References

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