Annotations

Asperger’s syndrome

In 1944 the Viennese paediatrician, Hans Asperger, described a group of children with what he called ‘autistic psychopathy of childhood’. He was in no doubt that this was a long lasting personality disorder, affecting predominantly boys, and he outlined its features as follows:

- Solitariness
- Abnormalities of gaze, expression, and gesture implying emotional contact with other people
- Insensitivity to social cues
- Lack of feeling for others sometimes amounting to callousness
- Oversensitivity and insensitivity
- ‘Autistic intelligence’: inventive rather than imitative, with specific interests in restricted fields such as chemistry, poisons, mathematics, or art which could lead to creative achievements
- Educational delays of all kinds
- Rage or tears in the face of pressure to conform. With age, social adaptation often improved and the work adjustment of gifted autistic psychopaths was good, but their basic personality features endured and intimate relationships remained impaired. Sexual interests were often meagre and occasionally deviant.

While Asperger considered the condition as possibly pre-schizophrenic, apparently only two of the 400 such cases he saw subsequently developed this illness. In every case one or more biological relatives were affected with the full or partial syndrome. In later years, Asperger drew a firm distinction between his ‘autistic psychopathy of childhood’ and Kanner’s early infantile autism and he described the children, who differed from any he had seen before, as ‘highly intelligent children with interesting peculiarities, yet never the less with behaviour so difficult that they were almost impossible to keep in family or school... They achieve the highest university professorships or become artists—yet their quirks and peculiarities will remain with them for life’.

Two developments in diagnostic practice have occurred since then. Lorna Wing and Digby Tantam studied groups of seriously impaired children and adults (of whom only two out of 60 had married and only one had been in continuous employment since leaving school) with the features Asperger had described and found that many of their patients had had the symptoms of early infantile autism or of autism beginning in later childhood. Wing described the salient features as a failure in two way social interaction, difficulty in verbal and non-verbal communication, and impaired imagination, and she coined the diagnostic label ‘Asperger’s syndrome’. This she held to be equivalent to high level autism (that is with normal intelligence and no gross early language delays). The concept of an ‘autistic spectrum’ derives from this work. Wing’s criteria were also used by Gillberg to clarify the similarities between Asperger’s syndrome and infantile autism.

The second development arose from the identification of a group of children referred to a psychiatric clinic who described themselves or were described by others as ‘loners’. They had the features of schizoid personality disorder as recorded in the older psychiatric literature (perhaps not altogether familiar to Asperger, the paediatrician). They too, while much less impaired than the cases reported by Wing and Tantam, were like the children Asperger had described. Their personality characteristics were very long lasting, and more recent studies showed three quarters of them to fulfil the American diagnostic criteria (Diagnostic and Statistical Manual of Mental Disorders, DSM-III) for schizotypal personality disorder which is part of the schizophrenia spectrum (S Wolff et al, to be published). The main, and enduring, features of these subjects were solitariness, impaired empathy and emotional detachment, increased sensitivity, at times with paranoid ideation, circumscribed interest patterns, and unusual styles of communication. In childhood, a very few resembled but were in fact never diagnosed as autistic children. A very few were electively mute, but most of them presented with the features of schizoid personality itself, with educational difficulties or with the kind of conduct or mixed conduct and emotional disorders usually regarded as secondary to adverse life experiences. Specific developmental delays, including delays of language related skills, were much commoner than in other referred children (S Wolff, to be published). The schizoids were predominantly boys (ratio 4:1), were of slightly above average IQ; and came from an upwardly skewed social class background. Referral took place during the school years and the main difficulty was often an apparently inexplicable failure to conform to the ordinary demands of school life. Aggressive outbursts and pathologi- cal lying were occasional features. With few exceptions these children were regarded as ‘awkward’ rather than handicapped.

Special school arrangements allowing for greater privacy and reduced pressure to conform were helpful, as was the...
acceptance of the children's difficulties as constitutional: neither wilful nor parent engendered. In later life, many of them had good work achievements and many married, but their overall work adjustment, their heterosexual adjustment, and their psychiatric state were worse than those of other referred children grown up (S Wolff, to be published).

These children resembled the schizotypal children described by Nagy and Szatmari, 31 36 and their difficulties certainly overlapped with those of the patients of Wing and Tantam. But whether they too should be included in an 'autistic spectrum' will depend on the results of more definitive genetic studies.

For this reason, the arguments put forward by Professor Cox in this issue against a global diagnosis of autism, 11 12 and in defence of a distinct diagnostic category of Asperger's syndrome, although based on Wing's rather narrow definition, are to be welcomed.

Dorothy Bishop, like other workers, considers Asperger's syndrome to overlap both with autism and with specific developmental language disorders, often diagnosed as congenital dysphasia. 13 A number of children with these disabilities also have the social impairments of Asperger/schizoid disorders. She suggests a two dimensional model for autistic type disorders, with impaired meaningful verbal communication on one axis and impaired social relationships with circumscribed interest patterns on the other. Asperger's syndrome, on this model, would overlap both with autism and with specific language disorders, but would be characterised by low scores on the language disorder dimension and high scores on the social impairment/circumscribed interests dimension.

Work on the diagnosis of these disorders is of theoretical interest, especially in disentangling the causal factors involved. It is also of great clinical importance. Children with severe forms of Asperger's syndrome will need the same services as well functioning autistic children. The more intelligent and less obviously handicapped Asperger/schizoid children require above all to be identified as constitutionally impaired. 14 Their special make up needs to be understood. They may also need special arrangements made, particularly at school, in the knowledge that in later life when pressures for conformity are less than during the school years, many will be able to find their social niche, and a few to develop their rather exceptional gifts.

SULA WOLFF

University Department of Psychiatry,
The Kennedy Tower, 
Royal Edinburgh Hospital, 
Morningside Park, 
Edinburgh E10 5HF

1 Asperger H. Die atistischen Psychopathen im Kindesalter. Archiv fuer Psychiatrie und Nervenkrankheiten 1944;177:76--77.

Possible aetiological factors in childhood leukaemia

Leukaemia accounts for over half of all cancers diagnosed in children, with one in 2000 developing the disease before their 15th birthday. 1 8--10 Despite controversial improvements in treatment over the last 20 years, about half of those who develop childhood leukaemia die within five years of diagnosis. Speculations about the causes of leukaemia in children abound, and several new and exciting theories have recently been suggested.

Ionising radiation

Exposure to ionising radiation is known to cause leukaemia. 14--16 Furthermore, the fetus and young child seem to be more susceptible to the effects of ionising radiation than the adult. 6--7 The controversial suggestion of Stewart et al, made in 1958, that prenatal exposure to x rays might increase the risk of childhood leukaemia has taken several decades to settle. It is now widely accepted that as much as 5% of all childhood leukaemias could be caused by diagnostic radiography of the mother's abdomen while the child is in utero. 7 8--10

Today, the possible contribution of natural radiation is a controversial topic. Estimates of the proportion of all childhood leukaemias attributable to background radiation range from a few percent to nearly all. 7 11--13 Assumptions about the relative effects of external gamma radiation and internal alpha particles on children of different ages and babies of different gestations alter these estimates appreciably. 7 11 Discussion has intensified recently after the report by Dr Henshaw and his colleagues demonstrating a correlation between indoor radon levels in different countries and the incidence of leukaemia and other cancers in children. 17 They suggest that a sizable proportion of all childhood leukaemias could be caused by exposure to radon gas in the home. These issues are now being vigorously investigated, but at this stage the aetiological relevance of natural background radiation is unclear.

More controversial still is the hypothesis that germ cell mutations induced by ionising radiation might increase the risk of leukaemia in the progeny of those exposed. This suggestion comes from the investigations of Professor Gardner and his colleagues of the raised incidence of childhood leukaemia near the Sellafield reprocessing plant in West Cumbria. 18 They found that exposure of fathers at work to comparatively low doses of external ionising radiation was associated with an increased risk of leukaemia in their offspring. The main finding of a sixfold to eightfold excess in the children of those most heavily exposed is, however, based on only four cases. As well as exposure of fathers at work to ionising radiation, other explanations are possible. 19--21 For example, external gamma radiation may not be the relevant exposure and exposure to work at other harmful agents may be involved. In particular, it has been hypothesised that radionuclides may accumulate in the male reproductive organs. Such a mechanism could explain both the high mortality from prostatic cancer found in highly exposed nuclear workers, and the increased incidence of leukaemia in the offspring of radiation workers at Sellafield. 16 17 Mothers' exposure to radiation before and during pregnancy may also be important, and could occur if