

Frederick Batten (1865-1918): father of paediatric neurology

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Abstract

Frederick Batten made many major contributions to neurology and rehabilitation. He was one of a group of eminent physicians at Queen Square in the Edwardian period and devoted his energies to paediatric neurology. Amongst many published works his papers on familial 'Cerebral degeneration with symmetrical changes in the maculae' (Batten's disease), subacute combined degeneration of the cord, and dystrophia myotonica are highlighted and summarised.

Key words

Paediatric neurology; Batten's disease; Neuronal Ceroid Lipofuscinoses; Subacute Combined Degeneration Of The Cord; Dystrophia Myotonica.

If you had passed through the portals of Queen Square in the Edwardian period spanning World War 1, you would have been confronted if not greeted by a group of eminent physicians. One such, often called the "father of paediatric neurology" was Frederick Batten*, who made many major contributions to neurology and rehabilitation.

Figure 1: Frederick Eustace Batten



He was born in Plymouth, son of JW Batten, Q.C, and his wife Sarah. He attended Westminster School and Trinity College, Cambridge, and graduated in medicine in 1891 from St Bartholomew's Hospital. He obtained his MD in 1895, and became FRCP in 1901. He was appointed Physician to the National Hospital for the Paralysed and Epileptic (pathologist, 1899; physician, 1900–1918; dean, 1908–1918) and to the Children's Hospital, Great Ormond Street. Both he served until his early death.

Amongst 106 published papers three are of particular interest: 1. Family cerebral degeneration with macular change: Batten's disease; 2. Subacute combined degeneration of the cord (SACD); and 3. Myotonia atrophica (Dystrophia myotonica).

Batten's disease

Batten described two cases in 1903,^{1,2} from the Hospital for Sick Children. The original description has been attributed to Otto Christian Stengel (1794-1890), who in 1826 described from Røros in Norway, a "singular illness" affecting four children of a local family.³ Interestingly Batten cited an earlier 1897 study⁴ on familial macula degeneration by his brother Rayner D Batten, but with hindsight there was no mental defect and no cerebral involvement. Frederick Batten described symmetrical changes in the maculae in two members of a family (see pedigree below) with cerebral degeneration starting about the age of six, now known as Batten's disease.

Batten's disease is a group of rare inherited autosomal recessive disorders that present in childhood with retinal and cerebral degeneration and terminate fatally. It is characterised by the intracellular accumulation of autofluorescent lipopigment storage material with different ultrastructural patterns. He described clinical features including visual impairment resulting in blindness; epilepsy; myoclonic jerks; impaired speech, language and swallowing; and a deterioration of motor skills that result in immobility. The afflicted child becomes totally dependent and death occurs in childhood or early adult life.

It is the most common form of the Neuronal Ceroid Lipofuscinoses (NCLs), linked to the CLN3 gene.⁵ Historically, the NCLs were classified by age of onset as infantile, late infantile,

juvenile or adult NCL. Juvenile NCL has also been called: Vogt-Spielmeyer disease, and Spielmeyer-Sjogren disease, but Batten's disease is often used generically. Each variant has differences in the rate of progression and clinical features. Treatment is of no avail save for cerliponase alfa (Brineura) which may slow progression of children with CLN2, the late infantile variant.

Batten's principal contribution was his recognition of the recently described neuronal storage diseases, and to realise his syndrome differed from Waren Tay-Sach's disease:

The difference lies in (1) the absence of race proclivity; (2) the absence of the characteristic macular change; (3) the difference of age. What the nature of the poison may be ... is a problem yet to be solved.⁶

Subacute combined degeneration of the cord (SACD)

Though described first by Leichtenstern in 1884,⁷ and by Putnam⁸ and Dana⁹ in 1891 the most comprehensive account was in a 70-page paper written by Batten with Risien Russell and Collier (Figure 2).¹⁰

SUBACUTE COMBINED DEGENERATION OF THE SPINAL CORD.

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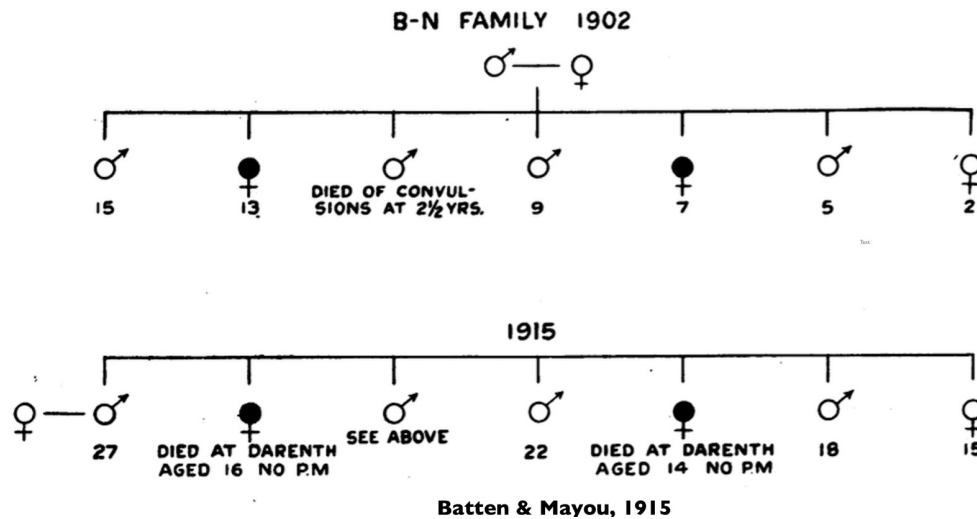
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INTRODUCTION.

MUCH as we feel the need of some other name under which to describe the affection, seven cases of which we are about to record, most of the papers that have appeared on the subject have borne the above or some similar title, so that to introduce a new name for the affection now would probably lead to confusion, however much it might serve to obtain for our own cases recognition that they are examples of a distinct morbid entity.

By combined degeneration of the spinal cord is meant an affection in which tracts of different function are concomitantly affected. It is, therefore, obvious that under such a title might legitimately be included a variety of different affections, each of which is characterised by the concomitant implication of tracts of different function. Examples of such affections are to be found in the "Ataxic Paraplegia" of Gowers, if there be such an affection distinct from the class of case with which we are more immediately concerned in our present paper; "Friedreich's Ataxy," "Cerebellar Heredo-Ataxy," such cases of "General

Figure 2. Russell JSR, Batten FE, Collier J. Subacute Combined Degeneration of the Spinal Cord. Brain 1900.



They described nine patients with autopsies in seven. The illness was in three stages of progressive paraplegia with ataxy and sensory loss in the lower limbs culminating in complete paraplegia; absent knee jerks; anaesthesia; wasting, and double incontinence. In respect of the recognised association with pernicious anaemia they noted:

‘... Some of the most typical cases presented no anaemia throughout the course ... others only late in the disease, while in other cases anaemia was an obtrusive symptom from the first and preceded the nervous symptoms by many months.’

They also observed that nomenclature was a problem:

‘A name other than “combined degeneration” would undoubtedly do much to establish the affection as a distinct morbid entity, ... in consequence of the fact that so many different diseases of the spinal cord are characterised by combined degeneration of tracts of different function.’

Leichtenstern, Putnam and Dana had all noted an association with anaemia, and Lichtheim specified pernicious anaemia.¹¹ Many years later were shown the causal cobalamin Vitamin B12 deficiency present in raw liver (Whipple Minot, Murphy, 1926, Nobel prize 1934¹²) and the associated deficiency of intrinsic factor needed for cobalamin absorption.

Dystrophia myotonica

Batten and Gibbs also provided one of the earliest detailed accounts of Dystrophia myotonica under the title of myotonia atrophica, published in 1909, the same year as Steinert’s paper.¹³ Charles Dana had described the syndrome in 1888.¹⁴ The salient features that Batten and Gibbs observed were:

‘...A group of cases which present the rare association of muscular atrophy with a slow relaxation of muscles after voluntary contraction. The muscular atrophy has a distribution which is peculiar and corresponds to none of the well known types of myopathy.’¹⁵

They recognised the disease as an entity distinct from Thomsen’s myotonia congenita and their paper includes the first photograph unmistakably portraying the disease. The association with cata-

tracts was not noted until Greenfield’s accounts of 1911 and 1923.^{16,17} It is now recognised as autosomal dominant DM1, with Cytogenetic locations: 19q13.32., typically showing: myotonia, myopathy, cataracts, hypogonadism, frontal balding, and ECG changes.

As a children’s physician, Batten ‘was in the first rank.’¹⁸ In 1913, with Sir Archibald Garrod and Thursfield he published a well-known textbook on Diseases of Children. In his Lumleian lectures at the Royal College of Physicians and re-published in Brain, June 1916, Batten detailed the features of poliomyelitis.¹⁹ His research was both clinical and pathological. He published several papers on progressive spinal muscular atrophy of infants (Werdnig-Hoffmann type) and on poliomyelitis (infantile paralysis); he devised corrective celluloid splints.^{20,21} Fittingly, in 1952 the intensive care unit for patients with respiratory paralysis at the National Hospital was named the Batten Unit in his honour.

His last communication was on epidemic stupor, jointly with his friend and renowned paediatrician, Sir George Frederick Still (1868-1941).²² Sir Gordon Holmes (1876–1965) recalled:

‘Batten’s approach was scientific...interpret[ing] symptoms as disturbances of function and determin[ing] the changes in structure ... to which they were due. His honesty, simplicity and directness impressed all who came in contact with him’.¹⁹

His students knew him as ‘Freddie’. He was described by TT Higgins as:

‘a brisk, lithe figure with a conspicuous domed head and lively eye, quick, tumbling speech ... and an intense interest in current affairs, but first and foremost concerned with the well-being of his patients and the parents, relatives, nurses and doctors who administered to them.’²³

Known as a perfectionist, disliking sloppiness and indecision, he nonetheless was considered lovable, modest, genial, and tolerant. Batten died aged only 52 from haemorrhage following surgery for prostatic obstruction.

*Amongst his colleagues were: T Buzzard, H Charlton Bastian, Sir David Ferrier, Sir WR Gowers, JA Ormerod, Howard H Tooth, Tames Taylor, JS Risien Russell, W Aldren Turner, JS Collier, E Farquhar Buzzard, T Grainger Stewart, Gordon M Holmes, CM Hinds Howell, and SA Kinnier Wilson.

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