Hebephrenia is dead, long live hebephrenia, or why Hecker and Chaslin were on to something

Alvaro Barrera, Owen Curwell-Parry & Marie-Claire Raphael

SUMMARY
Since its first description in 1863, ‘hebephrenia’ has highlighted a group of patients characterised by an early onset of illness, formal thought disorder, bizarre behaviour and incongruent emotional expression. A proportion of patients with the most severe form of mental illness have a clinical presentation that is best captured by this diagnosis. Here, we outline the construct of hebephrenia and two of its core overlapping constituent parts: bizarre behaviour and the disorganisation dimension. We argue that, despite the removal of hebephrenia (disorganised schizophrenia) from DSM-5, clinicians should consider it as a differential diagnosis, particularly in suspected personality disorder.

DECLARATION OF INTEREST
None.

KEYWORDS
Education and training; history of psychiatry; psychotic disorders; schizophrenia.

On hebephrenia
Hebephrenia’s description has remained fairly stable over time. As skilfully recounted by Kraam & Phillips (2012), hebephrenia was first described by Kalilbaum (1863), further elaborated by Hecker (1871) and Daraszkiewicz (1892) and clarified by Dide & Guiraud (1956); the common themes that resonate are those of early onset, bizarre behaviour and formal thought disorder. Remarkably, Hecker characterised hebephrenia by puerile, silly behaviour, often seemingly carried out only ‘to amuse or dupe others’, occurring in the absence of elevated mood and, with its aimlessness, pushing away or even irritating those who observe it. He even warned that individuals were likely to be mistaken as malingerers (Kraam 2009).

Hecker’s hebephrenia became the core of Kraepelin’s dementia praecox in the fourth edition of his textbook (1893), and it appeared as a subtype of dementia praecox in the fourth edition of Psychiatry: A Textbook for Students and Physicians (1896). A few years later, numerous examples of hebephrenia appeared throughout Bleuler’s influential monograph Dementia Praecox or the Group of Schizophrenias (Bleuler 1911), suggesting that for him hebephrenia and schizophrenia were almost coterminous.

A multifaceted description of hebephrenia was also provided by Leonhard (1965), who followed up over 1500 individuals with psychosis for long periods of time, in some cases decades. He classified the people diagnosed with schizophrenia into three groups: those with systematic schizophrenias, those with unsystematic schizophrenias and those suffering from cycloid psychoses. The systematic schizophrenias were characterised by an insidious onset, relentless course, poor prognosis, clear clinical delimitation and, curiously, no genetic influence (Franzek 1998). Hebephrenia, part of the systematic schizophrenias, had an early onset, emotional disintegration, episodes of severe excitement and aggression, and non-specific fleeting psychotic symptoms.

ICD and DSM classification
In ICD-10 (World Health Organization 1992) hebephrenia appears as hebephrenic schizophrenia, a condition that, in addition to meeting the generic criteria for schizophrenia, also presents with signs in the domains of affect, behaviour and thought. Affect shows sustained flattening, becoming shallow or inappropriate; behaviour can appear aimless, irresponsible and unpredictable, often with mannerisms; and thinking becomes disjointed, rambling or incoherent. There is also a tendency to social isolation and poor prognosis associated with the rapid development of negative features. Hebephrenia can only be diagnosed in adolescents or young adults and when hallucinations or delusions do not dominate the clinical picture.

A similar description of hebephrenia appeared in DSM-IV, as the subtype disorganised schizophrenia (American Psychiatric Association 1994). However, DSM-5 (American Psychiatric Association 2013)
has controversially eliminated disorganised schizophrenia (as well as the other subtypes of schizophrenia), a decision we regard as problematic for two reasons. First, psychiatrists will become less aware of hebephrenia and the fact that its recognition is based on signs (e.g. bizarre behaviour, inappropriate affect and formal thought disorder) in the relative absence of reported symptoms (e.g. hallucinations and delusions). This lack of familiarity with hebephrenia’s presentation might lead to people with a severe and enduring mental illness being diagnosed as having, for example, a personality disorder. Second, from a research point of view, subsuming hebephrenia into ‘generic’ schizophrenia, could leave a potentially useful phenotype prematurely abandoned.

An alternative proposal
A more radical approach is that of Taylor et al (2010), who suggest abandoning schizophrenia as a diagnosis and replacing it with hebephrenia. They argue that, although the current concept of schizophrenia encompasses a highly heterogeneous group of patients in terms of onset, course, biology and response to treatment, hebephrenia might actually provide a more homogeneous group for both clinical and research purposes. They propose the following diagnostic criteria for hebephrenia: (i) a specific prodrome during childhood, including cognitive, emotional, motor and socialisation difficulties, as well as occasional perceptual distortions; (ii) cognitive and motor deficits (e.g. poor sequential and fine-hand movements, dyspraxia and poor eye-tracking); and (iii) clinical features that include reduced emotional expression, avolition and apathy, formal thought disorder, delusions of passivity as well as sustained, clear voices perceived as originating from outside the person’s head. Although their rationale for including some of Schneider’s first-rank symptoms as part of hebephrenia is unclear, their proposal is open to empirical falsification.

Prevalence, prognosis and genetic risk
However we define it, hebephrenia remains a significant problem. The World Health Organization’s Ten Country Study found that it was present in 13% of cases of schizophrenia in ‘developed’ countries and 4% of cases occurring in ‘developing’ countries (Jablensky 1992), with its prevalence depending on the diagnostic criteria used (Stompe 2005). Regarding prognosis, Uggerby et al (2011) studied the Danish national register of all patients with schizophrenia, finding that hebephrenia was significantly associated with long-term institutionalisation (in-patient care), lower educational achievement, higher doses of antipsychotics and polypharmacy. Regarding genetic risk, Kendler et al (1998) reported that relatives of those with hebephrenia-like presentations showed a dramatically increased risk of schizophrenia, compared with relatives of those with other schizophrenia presentations.

On bizarre behaviour
Clinically, ‘bizarre behaviour’ refers to behaviour that strikes the observer as odd, without a clear motivation or goal and outside the person’s social and cultural norms, and that cannot be accounted for by other mental phenomena such as delusions, hallucinations, developmental disorders, mood disorders, substance misuse, dissociation or organic factors (e.g. frontal lobe syndrome).

Bizarre behaviour also has prognostic associations. For example, Owens et al (2010) found during 12-month follow-up of individuals with a first-episode of schizophrenia that bizarre behaviour and unemployed status independently increased the risk of relapse, with bizarre behaviour making the single biggest contribution. Similarly, Castle et al (1994) studied 484 first-contact patients with non-affective psychosis and found that bizarre behaviour, persecutory delusions and auditory hallucinations, as well as a diagnosis of schizoaffective disorder, were all more common among those admitted to hospital. Bizarre behaviour often makes people ‘stand out’ and it ranks highly in the lay view of schizophrenia and in its associated stigma (Rogers 2005, p. 28).

On the disorganisation dimension
Although hebephrenia is a categorical notion, the concept of disorganisation offers a useful dimensional approach to the same set of clinical phenomena, namely formal thought disorder, bizarre behaviour and incongruent affect. Indeed, since Liddle’s (1987) description, disorganisation has been identified as the third dimension of schizophrenia’s psychopathology, along with reality distortion (positive symptoms) and psychomotor poverty (negative symptoms). This has been demonstrated cross-sectionally, prospectively and in drug-naïve schizophrenia patients (Andreasen 1995; John 2003). Importantly, disorganisation also appears to be associated with poorer outcomes (Ortiz 2015).

The disorganisation dimension has clear neuro-psychological, anatomical and genetic associations. Regarding neuropsychological function, Ventura et al’s (2013) meta-analysis of 154 studies found a moderate association between disorganisation and global as well as specific cognitive dysfunctions, whereas the association between reality distortion
(delusions and hallucinations) and cognitive dysfunction was weak or non-existent.

Regarding functional anatomy, Goghari et al (2010) reviewed 25 functional magnetic resonance imaging (fMRI) studies and found that the disorganisation dimension was related to the function of the dorsolateral prefrontal cortex; negative symptoms were related to the function of the ventrolateral prefrontal cortex and ventral striatum; and positive symptoms were related to the function of the medial prefrontal cortex, amygdala, hippocampus and parahippocampal region. From a structural point of view, Collin et al (2012) studied the association between five schizophrenia symptom dimensions (negative, positive, disorganisation, mania and depression) and brain volume change over 5 years in 105 people with schizophrenia and 100 healthy comparison participants. Remarkably, only greater severity of the disorganisation dimension was associated with more pronounced decrease of total brain and cerebellar volume over time.

Finally, regarding genetic factors, Rietkerk et al’s (2008) meta-analysis found that the heritability of the disorganisation dimension in both twin and affected-sibling studies was more consistent in comparison with the heritability of the ‘reality distortion’ and ‘psychomotor poverty’ dimensions.

Explaining hebephrenia: discordance

Why do formal thought disorder, bizarre behaviour and incongruent affect significantly co-occur? The work of the French psychiatrist Philip Chaslin (1857–1923) on discordance might help to provide an answer. For Chaslin (Lantéri-Laura 1992), discordance was a kind of second-order phenomenon referring to the lack of the expected harmony between the gestures, emotions and content of a person’s statements: for example, when a patient puzzlingly smiles while angrily describing fears of being poisoned by his parents. Such discordance within or between action, affect and language would leave both patients and interlocutors with a sense of uneasiness. Of note, Chaslin believed that discordance increased the risk of psychotic disorders evolving into chronicity, not unlike the prognostic implications of hebephrenia and bizarre behaviour mentioned above.

Conclusions

We have outlined the construct of hebephrenia, including two of its core overlapping constituent parts, namely bizarre behaviour and the disorganisation dimension. The disorganisation dimension is associated with functional and structural neural dysfunction, cognitive impairment and significant heritability; bizarre behaviour appears to be associated with a relapsing course, hospital admission and stigma.

We urge clinicians to consider hebephrenia as a differential diagnosis in individuals who present with erratic and challenging behaviour, in the relative absence of symptoms such as hallucinations or delusions. It is worth keeping in mind the warning from early authors that people with hebephrenia may appear deliberately difficult or not genuine. The key here is the word ‘appear’. People with hebephrenia are unable to provide a reason as to why they have been abusive and disinhibited towards other people. They do not offer a comforting exculpatory explanation based on hallucinations or delusions. In today’s busy wards and clinics, where all too often feeling repelled or irritated by a patient translates into a crude countertransference diagnosis of personality disorder, these individuals run a risk of misdiagnosis.

References


Bleuler E (1911) Dementia Praecox oder Gruppe der Schizophrenien [Dementia Praecox or the Group of Schizophrenia]. Deuticke.


World Health Organization (1992) The ICD-10 Classification of Mental and Behavioural Disorders: Clinical Descriptions and Diagnostic Guidelines. WHO.