



## Agnosia for bird calls

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### ABSTRACT

The cognitive organisation of nonverbal auditory knowledge remains poorly defined. Deficits of environmental sound as well as word and visual object knowledge are well-recognised in semantic dementia. However, it is unclear how auditory cognition breaks down in this disorder and how this relates to deficits in other knowledge modalities. We had the opportunity to study a patient with a typical syndrome of semantic dementia who had extensive premorbid knowledge of birds, allowing us to assess the impact of the disease on the processing of auditory in relation to visual and verbal attributes of this specific knowledge category. We designed a novel neuropsychological test to probe knowledge of particular avian characteristics (size, behaviour [migratory or nonmigratory], habitat [whether or not primarily water-dwelling]) in the nonverbal auditory, visual and verbal modalities, based on a uniform two-alternative-forced-choice procedure. The patient's performance was compared to healthy older individuals of similar birding experience. We further compared his performance on this test of bird knowledge with his knowledge of familiar human voices and faces. Relative to healthy birder controls, the patient showed marked deficits of bird call and bird name knowledge but relatively preserved knowledge of avian visual attributes and retained knowledge of human voices and faces. In both the auditory and visual modalities, his knowledge of the avian characteristics of size and behaviour was intact whereas his knowledge of the associated characteristic of habitat was deficient. This case provides further evidence that nonverbal auditory knowledge has a fractionated organisation that can be differentially targeted in semantic dementia.

### 1. Introduction

The cognitive organisation of knowledge about nonverbal sounds remains poorly understood. This is attributable in part to a lack of detailed neuropsychological models of nonverbal auditory semantics and also the comparative rarity of reports of selective auditory agnosia, which might reveal the critical underlying cognitive architecture (Engelien et al., 1995; Clarke et al., 2000; Hattiangadi et al., 2005; Saygin et al., 2010; Slevc and Shell, 2015). It has been proposed that the processing of sounds as 'auditory objects' may be organised analogously to visual object processing, with corresponding neural mechanisms in auditory cortex and its connections in the temporal, parietal and frontal lobes (Goll et al., 2010a, 2010b; Brefczynski-Lewis and Lewis, 2017). However, opportunities to resolve key issues in auditory cognition – based on the study of patients with relevant deficits – remain limited.

One such important issue concerns the extent to which nonverbal auditory knowledge is differentiated. In the visual modality, it is relatively well established that object recognition is hierarchical (encompassing different levels of knowledge, ranging from general and

superordinate to more specific and fine-grained) and categorical (knowledge about different kinds of objects having cognitive and neural substrates that are at least partly separable) (Warrington, 1975; Jefferies and Lambon Ralph, 2006; Lambon Ralph et al., 2010; McCarthy and Warrington, 2016). The extent of differentiation within the visual semantic system is modulated by experience, as illustrated by the effects of brain damage in individuals possessing specific expertise with certain object categories (such as plants or cars: Jefferies et al., 2011). It is not clear whether similar cognitive organisational principles apply to auditory objects. In the case of one specialised semantic domain – knowledge about familiar people – impaired auditory recognition (phonagnosia) selective within the auditory modality and between modalities has been documented, to set alongside the better known visual equivalent of prosopagnosia (Hailstone et al., 2010, 2011; Luzzi et al., 2017). Specific agnosias have also been described for the equally specialised auditory domain of music (Ayotte et al., 2000; Clark et al., 2015). Studies of patients with auditory agnosia following focal brain damage have suggested that recognition of environmental sounds may dissociate from other kinds of auditory information processing

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(Engelien et al., 1995; Clarke et al., 2000; Hattiangadi et al., 2005; Saygin et al., 2010). However, a more fine-grained analysis of environmental sound recognition has remained largely elusive, in part because knowledge about sounds is generally not graded comparably with voices or melodies and often lacks a precise equivalent in other sensory modalities. Further key issues in semantic cognition concern the extent to which different input modalities contribute to multimodal or amodal conceptual representations and the representation of unique entities (Warrington, 1975; McCarthy and Warrington, 1988; Lambon Ralph et al., 2010; Wong and Gallate, 2012; Lambon Ralph, 2013; Gainotti, 2017). Here again, nonverbal sound is potentially an important role in the development of our current understanding of the human semantic system (Warrington, 1975; Lambon Ralph et al., 2010; Lambon Ralph, 2013; McCarthy and Warrington, 1988, 2016). Semantic dementia is a highly coherent clinic-anatomical syndrome; it typically presents as a fluent progressive aphasia led by loss of vocabulary but typically evolves to multimodal impairment of conceptual and object knowledge (Hodges and Patterson, 2007; Lambon Ralph, 2013), underpinned by selective disintegration of the neural networks that mediate semantic processing. These networks are centred on the anterior temporal lobes and semantic dementia is characteristically associated with anterior temporal lobe atrophy, usually more marked in the left cerebral hemisphere (Hodges and Patterson, 2007; Fletcher and Warren, 2011). Impaired recognition of sounds is well attested in SD (Bozeat et al., 2000; Goll et al., 2010a, 2010b; Hsieh et al., 2011; Hailstone et al., 2011; Golden et al., 2015). Indeed, SD constitutes a unique crucible for exploring the cognitive organisation of auditory and other nonverbal knowledge systems.

We recently had the opportunity to address these issues concerning the cognitive organisation of nonverbal sound knowledge in a patient, BA, with semantic dementia (SD) who possessed unusual premorbid expertise in the domain of bird knowledge. In addition to its resonance in clinical neurology as a leading focal brain degeneration of younger life (Hodges and Patterson, 2007; Warren et al., 2013), SD has played a pivotal role in the development of our current understanding of the human semantic system (Warrington, 1975; Lambon Ralph et al., 2010; Lambon Ralph, 2013; McCarthy and Warrington, 1988, 2016). Semantic dementia is a highly coherent clinic-anatomical syndrome; it typically presents as a fluent progressive aphasia led by loss of vocabulary but typically evolves to multimodal impairment of conceptual and object knowledge (Hodges and Patterson, 2007; Lambon Ralph, 2013), underpinned by selective disintegration of the neural networks that mediate semantic processing. These networks are centred on the anterior temporal lobes and semantic dementia is characteristically associated with anterior temporal lobe atrophy, usually more marked in the left cerebral hemisphere (Hodges and Patterson, 2007; Fletcher and Warren, 2011). Impaired recognition of sounds is well attested in SD (Bozeat et al., 2000; Goll et al., 2010a, 2010b; Hsieh et al., 2011; Hailstone et al., 2011; Golden et al., 2015). Indeed, SD constitutes a unique crucible for exploring the cognitive organisation of auditory and other nonverbal knowledge systems.

While the processing of voices and melodies in SD has been analysed at some length (Omar et al., 2010; Hailstone et al., 2011; Hsieh et al., 2011; Golden et al., 2015), the organisation of environmental sound processing in this disorder has not been delineated in like detail. BA's special knowledge of birds is therefore particularly apposite: birds constitute a semantic domain that is finely graded (comprising a multiplicity of species with defining features superimposed on basic superordinate avian characteristics) and comparably accessible via the auditory and visual sensory modalities. The impact of semantic dementia on bird recognition should therefore expose the cognitive organisation of a highly differentiated category of environmental sound knowledge, and allow direct comparisons between knowledge modalities and with other specialised knowledge domains (such as human voices). Moreover, experienced bird enthusiasts ('birders') generally acquire knowledge of bird appearance and bird calls in tandem; it should therefore be possible to disambiguate the effects of acquired expertise from intrinsic modality-specific contributions to this knowledge category. In studying BA, we designed a neuropsychological experiment to probe different dimensions of avian knowledge relating to avian physical features and associated ethological characteristics, in parallel via auditory (bird call), visual (bird appearance) and verbal (bird name) input modalities. We created novel stimuli to assess knowledge of the same bird attributes via each of these input channels independently: conventional tests of semantic processing often rely on cross-modal matching tasks that involve two or more processing channels (e.g., names and pictures), however such tests confound interpretation of modality-specific effects. BA's performance on our novel semantic test was referenced to a control group of healthy older individuals with similar birding experience. In addition, we sought to compare BA's knowledge of birds with his knowledge of familiar people

via their voices and faces, in order to assess the specificity of any avian semantic deficit.

## 2. Methods

### 2.1. Clinical details of patient BA

BA is a right-handed male former credit union executive aged 65 who presented with a six year history of progressive loss of vocabulary. He had particular difficulty finding the names of people and objects and would ask the meaning of words such as 'spatial'. His conversation had become increasingly imprecise but remained fluent, with no history of speech sound errors or dysarthria. He was less inclined to read newspapers and magazines due to difficulty understanding their text. His general intellect was well preserved, including memory for recent autobiographical events and facility with financial transactions and household gadgets. His family considered that he had become a little more emotionally labile but there had been no significant behavioural or personality change and in particular, no instances of social disinhibition or faux pas. There was no past medical history of note nor any relevant family history. He had undergone pure tone audiometry on account of his impaired speech comprehension; this revealed only mild age-related hearing loss.

BA is a dedicated amateur birder with some 30 years' experience, including around 10 weeks each spring spent in birdwatching expeditions and over the years had also regularly attended courses in bird call recognition, visual identification and bird behaviour. He had extensive exposure to a range of bird species representing all major regions and habitats of the British Isles. He had noted waning of his ability to name birds or identify them from their calls over a similar timeframe to his evolving difficulty with general vocabulary. At the time of assessment, he was also becoming less competent at identifying birds visually but he continued to enjoy recognising and feeding the birds that visited his garden. There had been no suggestion of any difficulty recognising familiar faces or household items nor any difficulty recognising the voices of telephone callers or everyday noises. There had been no evident change in BA's appreciation of music.

On examination his speech was circumlocutory with impoverished content but grammatically correct and normally articulated. He exhibited impaired picture naming and single word comprehension; repetition of polysyllabic words and phrases, arithmetic, praxis and visuo-perceptual functions were intact. General neuropsychological assessment supported the bedside impression of severe anomia underpinned by a selective, primary semantic memory deficit, with associated deficits of episodic verbal and face memory but intact speech production, sentence processing, executive skills and auditory and visual perceptual functions in the context of very superior performance IQ (Table 1). The general neurological examination was normal.

Brain MRI (Fig. 1) showed asymmetric atrophy predominantly affecting the anterior, mesial and inferior temporal lobes, more marked on the left. Together with the clinical presentation and neuropsychological findings, the MRI appearances were typical of SD, as defined in current consensus diagnostic criteria (Gorno Tempini et al., 2011).

### 2.2. Healthy control participants

Three older amateur birders with no history of neurological or otological disease also participated in the experimental study. These healthy control participants were members of BA's birding group and had similar birding experience, such as attending the same fieldtrips and educational activities; their details are summarised in Table 2.

The study was approved by the local institutional ethics committee and all participants gave informed consent in accordance with the guidelines of the Declaration of Helsinki.

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