Teaching Course 16

Higher cortical function in neurology - an update - Level 2

Executive function and behaviour

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What are executive functions?
The term ‘executive function’ refers to brain processes that are deployed when behaviour needs to be ‘controlled’ or ‘supervised’.1,2

1. When the situation is novel or difficult
   Control or supervision might be necessary because the situation is novel or difficult. Under these circumstances, there might not be an automatic, habitual or routine response to the problem. Or the automatic response might be inappropriate. Take for example, the situation when the telephone rings in someone else’s house. Normally you would pick up the phone in your own house. That would be the automatic response, but in this context that would be inappropriate. You exert control to stop yourself responding in the automatic manner, and you also need to monitor whether the behaviour you selected was the appropriate response in any particular context.

2. When several cognitive processes need to be co-ordinated
   We often have to plan ahead and co-ordinate a sequence of actions correctly to achieve a goal or several goals. For example, you might need to plan your day in order most effectively to obtain items from a shopping trip which involves visiting several shops, drop children off at a party, deposit the laundry, visit a friend, give the car a wash and pick up the children on time, given likely local traffic conditions.

3. When we need to shift from the current behaviour to another one
   We also often need to multi-task and have to switch flexibly and rapidly from one behavioural ‘set’ to another, and back again, to achieve our goals. Imagine you are at your desk answering an important email when the phone rings. You know the call is urgent so you have take it, but you also need to complete your email within the next five minutes, so you try to both things properly, switching your attention from one to the other. You seem to be doing well but just then there’s a knock on the door. It’s a colleague who wants advice on a difficult case. You have to ask the caller on the phone to hold while you deal with this. Under these circumstances, being able to switch effectively from one behaviour to another requires control which most of us would find challenging, but possible.

Examples of processes that are considered to be executive functions are shown in Table 1.
Table 1 | Examples of executive functions

- Initiation of action
- Maintain / sustain / invigorate / energise behaviour
- Stop ongoing action
- Inhibit inappropriate behavior or prepotent responses
- Monitor consequences of behavior – error monitoring
- Switch to a different behavioural set – set shifting
- Exert control to minimize effects of conflict
- Planning ahead and prioritization
- Multi-tasking
- Working memory: manipulation of items in short-term memory
- Social and emotional control

One highly influential model of executive control was developed by Norman and Shallice. In their view, a ‘Supervisory system’ needs to control behaviour when it is non-routine, when we need to exert control or shift flexibly to alternative actions (Figure 1).

Figure 1 | Supervisory model of executive functions

In this model, under routine situations, behaviour is controlled by schemas which are triggered by familiar sensory information. However, in non-routine situations control needs to be exerted so the routine behaviour is not triggered. That control is exerted by the ‘Supervisory system’. This model was first developed by Norman and Shallice. Figure adapted from ref 1.
Frontal lobes, executive functions and the ‘dysexecutive syndrome’

Traditionally, the frontal lobes have been associated with executive functions.\(^1-^3\) Lesions to or atrophy of frontal regions can lead to a wide range of cognitive impairments and behavioural states. It is now appreciated that while some patients might manifest behavioural changes, sometime referred to as a ‘dysexecutive syndrome’ (Table 2) they may not show deficits on neuropsychological tests of executive function. Conversely, other patients can be impaired on executive function tests but show no evidence of behavioural disturbance.\(^4\)

<table>
<thead>
<tr>
<th>Behavioural changes associated with the dysexecutive syndrome(^4)</th>
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<tbody>
<tr>
<td><strong>Highly suggestive</strong></td>
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<tr>
<td>▪ Global hypoactivity with apathy and/or abulia</td>
</tr>
<tr>
<td>▪ Global hyperactivity with distractibility and/or psychomotor instability</td>
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<tr>
<td>▪ Stereotyped and perseverative behaviour</td>
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<td>▪ Environmental dependency – imitation and utilization behavior</td>
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<td><strong>Supportive deficits</strong></td>
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<tr>
<td>▪ Disorders of emotional control – apathy, euphoria, emotional lability</td>
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<td>▪ Disorders of social behaviour</td>
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<tr>
<td>▪ Disorders of sexual, eating and urinary behaviour</td>
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<td>▪ Spontaneous confabulation</td>
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<td>▪ Anosognosia</td>
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Many different pathologies affecting particularly the frontal lobes can lead to executive function deficits and/or the dysexecutive syndrome. These include focal lesions such as stroke; traumatic brain injury; neurodegenerative conditions including Alzheimer’s disease, vascular dementia, frontotemporal dementia (FTD), Parkinson’s disease dementia and Lewy body dementia; inflammatory disorders such as multiple sclerosis; developmental conditions such as attention deficit hyperactivity disorder; and psychiatric disorders such as schizophrenia.

Although executive function deficits and the dysexecutive syndrome are dissociable, there is often a strong correlation them. A recent large study in patients with behavioural variant FTD, primary progressive aphasia and corticobasal syndrome measured both executive function performance and behaviour associated with the dysexecutive syndrome.\(^5\) In addition, regions of atrophy associated with both executive function and the dysexecutive syndrome were mapped. The results show
regions unique to either, as well as regions of overlap (Figure 2). Note that deficits on EF tests can occur with atrophy to parietal regions as well as classical frontal ones.

**Figure 2 |** Regions of atrophy associated with executive function (EF) deficit, the dysexecutive syndrome (DS) or both. Areas in red represent regions uniquely associated with EF deficit. Note these include parietal regions as well as frontal ones. Blue areas are those associated uniquely with DS. Area in yellow denote those regions associated with both. Figure adapted from ref 5.

**How do we measure executive functions?**
A wide range of cognitive tests have been developed to measure executive functions in the clinic. Different centres use different batteries of such tests. One factor that constrains the selection is time available for a neuropsychologist to perform testing. Some neurologists have therefore elected to develop short test batteries that can be performed at the bedside.

Below are some widely used tests. Some of them can be performed without special material by a neurologist. It is important to appreciate though that *impairment on an executive function test does not necessarily mean that a patient has a deficit in executive function*. She could fail a test because she has language, visuospatial impairment or limb impairments that affect her ability to perform the test. Hence it is important not to test executive functions in isolation but to have a broader view of the cognitive profile of the patient. Interpretation has to be conducted in context.

**Fluency tests**
Fluency tasks probe the ability to generate as many items as they can within a fixed time. They can be performed easily in the clinic. *Verbal fluency* tasks require patients...
to generate within a minute as many words as they can beginning with a particular letter in the alphabet (e.g. letter ‘F’), excluding names. Non-verbal or design fluency tasks require patients to draw as many patterns as they can within a fixed time. The pattern might be constrained to joining up a series of dots in as many different ways as possible.

**Go-NoGo**

These tests assess the ability to withhold or inhibit an action when it is not appropriate. For example, a patient might be asked to tap the table when the examiner taps once, but inhibit any response when the examiner taps twice.

**Stroop test**

In this test, a patient must withhold the prepotent or routine response. They are asked first to read a list of words which are lists of colours (blue, red, etc.) which are printed in different colour inks. This is usually easy for all of us. The second stage requires the patient to name the ink colour in which a word is written. This is harder because we have to withhold the urge to simply name the colour word instead of the ink colour.

**Luria’s manual sequence**

In this bedside test, the examiner, seated in front of the patient, performs three times with their left hand the manual sequence devised by Luria which consist of fist–edge–palm. The patient is then asked to continue repeating this sequence.

**Trail making**

This is simple bedside test that requires patients to switch between one set of stimuli and another. In the baseline (Trails A) test, patients are asked simply to join up (connect by drawing lines) consecutively numbered circles: 1-2-3-4-5-6 etc. In the Trails B test they are asked to switch between two stimulus sets (letters and numbers) joining them up in the right order: 1-A-2-B-3-C etc.

**Wisconsin card sort**

This is a classic test which has been used to probe abstraction, concept formation and behavioural flexibility. The patient is asked how to show how they might sort cards by a particular rule. They are first asked to place cards one by one next to four stimulus cards (e.g., one with a red triangle, one with 2 green stars, another with 3 yellow crosses and another with four blue circles). They have to deduce the rule that is being applied by the examiner to sort the cards, e.g. this might first be by colour. After a run of ten correct runs, the examiner changes the rule, e.g. to sorting by shape, and the patient must deduce the rule has changed and switch to using the new principle.
**Tower of London task**\textsuperscript{6,14}  
Tower tasks require patients to apply strategies to solve problems of increasing difficulty. The patient is asked to move a series of coloured balls on three sticks to achieve a goal position, moving only one ring at a time.

Several other tests have been developed to produce more ecologically valid tests because some patients who have obvious problems at work or home can pass many standard tests such as those described above. In the **Multiple errands test**\textsuperscript{15–16} participants are taken shopping and asked to complete several different errands in any order they choose, while in the **Six elements test**\textsuperscript{13} they are asked to complete six tasks (two sets of three different tests) in 15 minutes with the aim of maximizing their total score.

**How do we assess the dysexecutive syndrome?**  
A great deal of information can be obtained from the clinical interview: by observation of the patient, by taking a history from her alone and, most importantly, by obtaining an independent history from the informant. Five minutes spent alone with the carer can reveal difficulties that would not always be apparent by conducting an interview with both the patient and carer present. Some structured instruments have also been developed\textsuperscript{4,17}, including the Frontal Systems Behaviour Scale (FrSBe)\textsuperscript{17,18} which is a 46-item rating scale with two forms: one to be completed by the patient and the other by the carer.

In this teaching course lecture, we’ll review executive functions and the dysexecutive syndrome, including using some published case histories as useful clinical examples.\textsuperscript{19–21}
References