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15 minute consultation: Tics and Tourette syndrome

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ABSTRACT

Tic disorders including Tourette syndrome (TS) are neuropsychiatric disorders that are common referrals to paediatricians, paediatric neurologists and child psychiatrists. Although differentiating tics and TS from other movement disorders is not difficult, it is essential to detect comorbid conditions and their contribution to TS.

CASE SCENARIO

Eight-year-old Tom was referred by his general practitioner (GP) asking whether his movements and behaviour were tics. His parents reported noticing intermittent facial grimacing movements and grunting for the last six months. They report that it happens both at home and in school and was worse a few weeks back when they had consulted the GP. The GP advised that the episodes should be ignored, and since then they have been less frequent.

WHAT ARE TICS?

Tics are sudden recurrent individually recognisable vocalisations, intermittent movements or movement fragments that are almost always briefly suppressible and are usually associated with awareness of an urge to perform the movement.[1] Suppression of a tic usually leads to growing discomfort, which is alleviated by performing the tic. Characteristic features of tics are predictability of movement, predictability of onset and triggered by suggestion, sensory cues, stress, excitement or demonstration.[1 2] Tourette syndrome (TS) is in the spectrum of tic disorders but needs to fulfil certain diagnostic criteria (see below).

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TYPES OF TICS

Tics can be classified by description (table 1) and by pathophysiology (box 1). Pathologic tics can be further classified into primary sporadic, primary inherited and secondary.

DIAGNOSTIC CRITERIA AND SPECTRUM OF TIC DISORDERS INCLUDING TS

Tic disorders can be diagnosed using the Diagnostic and Statistical Manual of Mental Disorders V (DSM-V)[4] or the International Classification of Disease (ICD)-10[5] criteria and be classified as:

- provisional tic disorder (DSM-V) or transient tic disorder (ICD-10)—tics that occur on most days with a duration of 4 weeks to a year, not due to TS;
- chronic/persistent motor or vocal tic disorder—tics that occur on most days for longer than a year, not due to TS;
- TS—two or more motor tics and at least one vocal tic occurring for at least a year.

All the above must start before 18 years of age and not be due to physical conditions or medication side effect. There should not be a tic-free period of >3 months in a year.

PREVALENCE AND NATURAL HISTORY

Tics are present in up to 5% of the population, with a lifetime prevalence of ≥20% among boys.[6] The prevalence of TS is 7.7 per 1000 children (95% CI 3.9 to 1.51).[7] Motor tics usually begin between the ages of 3 and 8, and phonic/vocal tics usually follow the onset of motor tics after several years.[8] Uncomplicated tics peak in the early second decade with a marked reduction in tic severity by the age of 19 or 20 years (figure 1).[9]

The onset of TS is typically between 2 and 15 years, average of 6.4 years, and is manifested in 96% of patients by 11 years of age.[10] The disorder is usually diagnosed 2–3 years after onset of tics. Peak severity of tics are usually at 9–11 years of age. Five to ten per cent of patients have an intensifying course without improvement. In about 85% of patients, however, symptoms diminish during or after adolescence[11] (figure 2).

Table 1 Classification of tics by description

Motor	
Simple	Brief, meaningless, abrupt movement fragments (eg, eye blinking)
Complex	Longer, involving more muscle groups, more goal-directed in character (eg, scratching, chewing)
Phonic/vocal	
Simple	Fast meaningless sounds or noises (eg, sniffing, throat clearing, grunting, barks, high-pitched squeaks)
Complex	Syllables, words, phrases, odd patterns of speech, echolalia, palilalia and coprolalia

Tics in TS affects more males than females (4.3:1).[12] They are characteristically waxing and waning.[2] Frequency and severity of tics in childhood is hardly predictive of tic severity in adulthood. Tics and TS are associated with other neurobehavioural conditions, particularly attention-deficit hyperactivity disorder (ADHD) and obsessive-compulsive disorder (OCD) (figure 1).

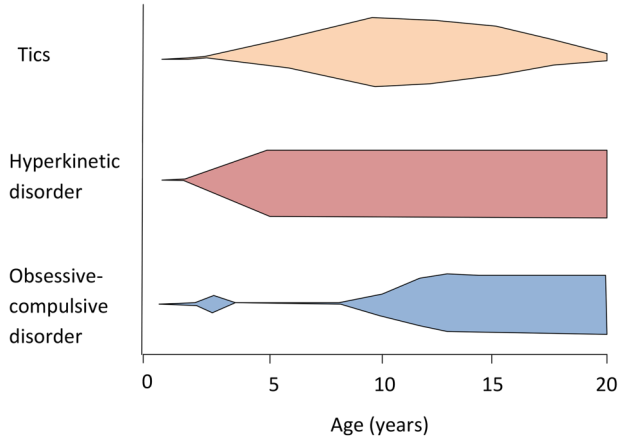

Figure 1 Relationship between age and severity of tics and coexisting disorders.

PATHOPHYSIOLOGY

The disorder likely results from a disturbance in the striatal–thalamic–cortical (mesolimbic) spinal system, in which dopamine plays an important role, which leads to disinhibition of the motor and limbic system.[13] Alterations in the cortico–striato–thalamo–cortical circuits in patients with TS have been found on MRI and electrophysiological studies.[14] Family studies strongly indicate a genetic component in TS, with shared genetic influences between tics and OCD. The link to ADHD, however, is unclear.[14] Several loci have been identified as candidate susceptibility regions, and mutations in SLITRK1 gene[15] and HDC gene[16] appear to be a rare cause of TS.

Box 1 Classification of tics by pathophysiology

Physiologic tics
Motor mannerisms[3]
- Repetitive, distinctive behavioural trait that does not interfere with life or cause self-injury (eg, brushing coat repetitively, persistent hair twirling)

Pathologic tics
Primary
- Sporadic
- Provisional/transient tic disorder (<1 year)
- Chronic/persistent motor or vocal tic disorder (>1 year)
- Adult-onset (recurrent) tics
- Tourette syndrome
- Inherited
- Tourette syndrome
- Huntington's disease
- Primary dystonia
- Neuroacanthocytosis

Secondary ('Tourettism')
Infections: encephalitis, Creutzfeldt–Jakob disease, post-streptococcal disorders
Drugs: stimulants, levodopa, carbamazepine, phenytoin, phenobarbital, antipsychotics (tardive tics)
Toxins: carbon monoxide
Developmental: pervasive developmental disorders
Brain injury: head trauma, stroke
Genetic: neurocutaneous syndromes, chromosomal abnormalities, neuroacanthocytosis
Psychiatry: schizophrenia, psychogenic tics

DIFFERENTIAL DIAGNOSES

Other hyperkinetic movement disorders (dystonia, choreoathetois, myoclonus, tremor, stereotypy)

Dystonia is usually triggered by and interferes with voluntary movement, but tics are usually suppressed by and do not interfere with voluntary movement.[1] Tics can be differentiated from athetosis, chorea and myoclonus by the lack of continuity of the movement, the intervening periods of normal movement and the lack of interference with ongoing tasks. Tics are predictable and repeatable compared with chorea and myoclonus. The clear initiation and termination of each individual tic movement and the lack of rhythmicity in timing of initiation of movement distinguishes it from tremor and stereotypies.[1] Unlike tics,

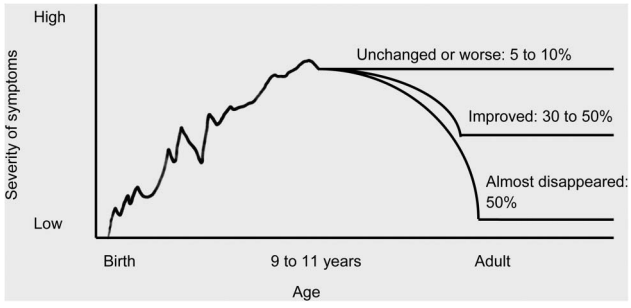

Figure 2 The clinical course of Tourette syndrome.

Table 2 Key features of hyperkinetic movement disorders

	Rhythmic	Repeated posture	Repeated stereotyped movement	Suppressible
Tics	No	Yes	Yes	Usually
Dystonia	Rarely	Yes	Sometimes	Partial or only briefly
Chorea	No	No	Rarely	No
Athetosis	No	No	No	No
Myoclonus	Sometimes	Sometimes	Usually	No
Tremor	Yes	No	Yes	Sometimes briefly
Stereotypies	Yes	Sometimes	Yes	Yes (but young age makes it difficult to tell)

Table 3 Clinical differences of tic disorders and obsessive-compulsive disorders

Tics	Obsessive-compulsive disorder
Sudden, short (jerking)	Ritualised
Fragmented movements	Goal-directed behaviour (to reduce distress and/or prevent a feared event)
Sensorimotor urges	Thoughts/imaginations (obsessions)
Involuntary (clustered sequence)	Voluntary (cyclic)
Onset in primary school (one peak)	Onset pre-primary and post-primary school (two peaks). See figure 1
Waxing and waning (from seconds to months)	Little changes over time
Can occur in sleep	Never during sleep
Comorbid with ADHD, OCD, depression	Comorbid with anxiety disorders, eating disorders, TS, depression

ADHD, attention-deficit hyperactivity disorder; OCD, obsessive-compulsive disorder; TS, Tourette syndrome.

stereotypies usually start before the age of 2 years. Stereotypies are repetitive simple movements, vocalisation or behaviour such as head banging, waving or flapping that are present for >4 weeks and lack premonitory urge.[3] Table 2 outlines the key features of various movement disorders.

Obsessive-compulsive disorder

It can be difficult differentiating a tic from obsessive-compulsive behaviour (table 3) as they share many similarities and are indeed often comorbid. Both decrease with concentration, increase with emotional excitement and are suppressible (tics for a shorter term than OCD).[14]

Other conditions that mimic tics

Tics can also be mistaken for other conditions such as epileptic seizures, sleep disorders, attention problems and functional disorders (table 4).

COMORBIDITIES

TS is commonly associated with comorbidities (table 5).[12] Motor and vocal manifestations are more frequent in boys, whereas girls are more likely to have behavioural problems such as OCD.

ADHD symptoms in TS occur in the majority of cases before tic onset (figure 1). In our personal experience, we found that comorbid ADHD, when present, occurred in nearly 70% of affected children before the onset of TS. ADHD symptoms tend to decrease in 20% of children during adolescence, usually following a decrease in tics. ADHD in patients with TS are associated with increased irritability and

Table 4 Clinical conditions that could be mistaken for tics and vice versa

Tic phenomena	Differential diagnoses
Eye rolling	Absences
Blinking	Allergy, poor vision
Focussing on tic control	Attention problems
'Excessive' tics	Imitation/somatisation
Tripping	Medication-induced akathisia, juvenile parkinsonism
Neck jerking	Dystonia, medication-induced dyskinesia
Convulsive grimacing	Facial spasms or blepharospasms
Sniffing	Allergy, hay fever, cold
Scratching	Scabies, lice, allergies
Tics during sleep	Restless leg syndrome/benign epilepsy of childhood with centro-temporal spikes/parasomnias, sleep myoclonus

Table 5 Comorbid conditions in patients with Tourette syndrome

Attention-deficit hyperactivity disorder	60%
Obsessive-compulsive behaviour	32%
Obsessive-compulsive disorder	27%
Learning disorder	23%
Conduct disorder/oppositional defiant disorder	15%

rage attacks, increased vulnerability for drug abuse, depression and antisocial behaviour. TS plus ADHD appears to be a more severe condition than ADHD alone.[17]

Comorbidity of TS with OCD can make distinction between complex motor tics and compulsions difficult.[1] Comorbid obsessive-compulsive symptoms start at a later age (around 10 years) than tics (figure 1).

Table 6 Assessment of tics

History	
Tic history (salient points)	
Age of onset of first tics	To determine type of tic
Course and age at worst tic severity	To prognosticate progression
Determine most debilitating complaints and symptoms	To determine management
Is the movement suppressible?	Identifies if it is indeed a tic
Triggers, exacerbating and relieving factors	Identifies if it is indeed a tic
Fluctuation of symptoms	Identifies if it is indeed a tic
Circadian profile of tic activity (including sleep)	Differentiates from other movement disorders
Possible relationship between infections (throat and ear) with tic exacerbation	Consider streptococcal autoimmunity
Developmental history	Identify other possible behavioural and neurological conditions, particularly comorbid conditions (eg, ADHD and OCD)
Past medical history	
Medication—current and past	Identify medication-induced movement disorder
Family and social history	
Family functioning—parental coping styles, parental conflict, social network, financial and housing situation	Identify potential stressors and triggers
Parent and patient rating scales, eg, Strengths and Difficulties Questionnaire	Identify functional difficulties
Impact on tics on family, learning, quality of life	Identify potential stressors and triggers
Family psychosocial and medical history	Identifies psychiatric and/or neurological conditions in relatives particularly of tics, OCD and ADHD in first-degree family
Other	
Collaborative data (eg, family members, school, video of tics)	To corroborate information, identifies if it is indeed a tic as may not be observed in clinic
Examination	
General examination	
Dysmorphic features	Identify genetic syndromes particularly in association with learning difficulties/ autism spectrum. Consult clinical genetics and consider CGH-array if available.
Neurological examination	Exclude severe or progressive neurological disorders
Observation of tic	Differentiate from other movement disorders (table 2)
Fine motor skills, eg, writing and putting lid on pen	Differentiate from other movement disorders. Tics less pronounced when concentrating.
Investigations	
EEG, neuroimaging, laboratory studies	These are rarely indicated. They may assist in differential diagnoses when the presentation is not typical or deterioration is severe
Neuropsychological evaluation (intellectual function, academic attainments, motor skills, attention, executive function and memory)	If the child has comorbid ADHD or OCD
Yale Global Tic Severity Scale[18]	Measures likelihood of having TS

ADHD, attention-deficit hyperactivity disorder; CGH comparative genomic hybridisation; OCD, obsessive-compulsive disorder; TS, Tourette syndrome.

Normal obsessive-compulsive-like symptoms are present in many young children, peaking at 2.5 years of age. This disorder, when associated with tics, generally has a pre-pubertal age of onset. OCD can lead to periods of depression. In our personal experience, we found OCD occurred in 50% of children with TS. Comorbid OCD and ADHD are associated with poorer psychosocial functioning.[14]

ASSESSMENT

Salient features of clinical assessment and investigations are outlined in table 6. Obtaining a video recording of the episode on a camera or a mobile phone is most useful in making the correct diagnosis.

MANAGEMENT (SEE ALGORITHM) (figure 3)

Most children with simple motor tics do not require treatment. Knowledge of the temporal patterning of tics is fundamental for clinicians as it informs decisions about when to initiate or change management and when to 'watch and wait' after psycho-education and reassurance.[2]

Non-pharmacological and/or pharmacological interventions should be considered in addition to psycho-education for children with clear impairment associated with the tics[19] (box 2).

In the presence of comorbid conditions (ADHD, OCD, depression), it is better to treat the comorbidities first, as successful treatment of them will often diminish tic severity.[9]

NON-PHARMACOLOGICAL

Behavioural interventions

Habit reversal training (HRT) and exposure with response prevention (ERP) have the most evidence among the behavioural treatments for tics[20] and are considered first line. Treatments that are considered second line or 'add-on' are contingency management, function-based interventions and relaxation training. HRT offers a set of techniques to increase patients' awareness of occurrence of a tic, followed by a response training to inhibit the tic. In ERP, tics are viewed as conditioned responses to stimuli. By confronting the sensation, the stimuli evokes (exposure) and resisting the tic (response prevention) patients might learn to tolerate the sensation (habituation).

One of the oldest described behavioural treatment, massed (negative) practice or voluntary 'over-ticcing', involves repeated, rapid, voluntary performance of the tic for specified periods of time interspersed with brief periods of rest with an expectation that this may reduce tic frequency at other times. Current evidence suggests that this is of limited therapeutic value.[20] There is also no evidence that 'masking' tics help. Masking may not provide the same satisfaction as performing the tic unmasked and potentially could increase tic frequency in some children.

Psychosocial interventions

Psychosocial interventions include psycho-education, group work, information and support from voluntary organisations. Psycho-education in TS aims to improve the tolerance for symptoms and to support stress reduction. Psycho-education involves providing information about the long-term and short-term variability of tics, about the natural course and about possible coexisting problems. Psycho-education alone may be used in families who do not engage with or need more comprehensive treatments due to very mild tics or cannot access treatments due to lack of services.[20] Other psychosocial interventions are used as adjuncts to other therapies.

Box 2 Indication for treatment of tics

- Tics cause subjective discomfort (eg, pain and/or injury)
- Tics cause sustained social problems for the patient (eg, social isolation or bullying)
- Tics cause social and emotional problems for the patient (eg, reactive depressive symptoms)
- Tics cause functional interference (eg, impairment of academic progress)

PHARMACOTHERAPY

Pharmacotherapy should only be instituted by clinicians or units who have experience of managing tic disorders and of using the medication itself. These medications are not commonly used by most paediatricians and caution needs to be exercised as their use can be associated with significant side effects. In most instances where medication is being considered, it is preferable to refer the child to a specialist unit and an experienced clinician.

There is a paucity of studies directly comparing efficacy and safety of pharmacological agents, hence no firm recommendations can be made. Pharmacotherapy probably has the fastest onset of response compared with behavioural treatments, but this experience has never been tested in a clinical trial. In our personal practice, indications for pharmacological treatment are subjective discomfort, social isolation with bullying and harassment, reactive depressive symptoms and impairment of school performance. We use non-neuroleptics like clonidine as a first-line medication. We use neuroleptics as second line when non-pharmacological treatment and first-line treatment have been unsuccessful. As tics are thought to involve the dopaminergic pathway, modulating this, particularly by blocking the post-synaptic D2-receptors, is the main action of pharmacotherapy in tics.[19] Medication choice, doses and level of evidence of effectiveness are provided in table 7.

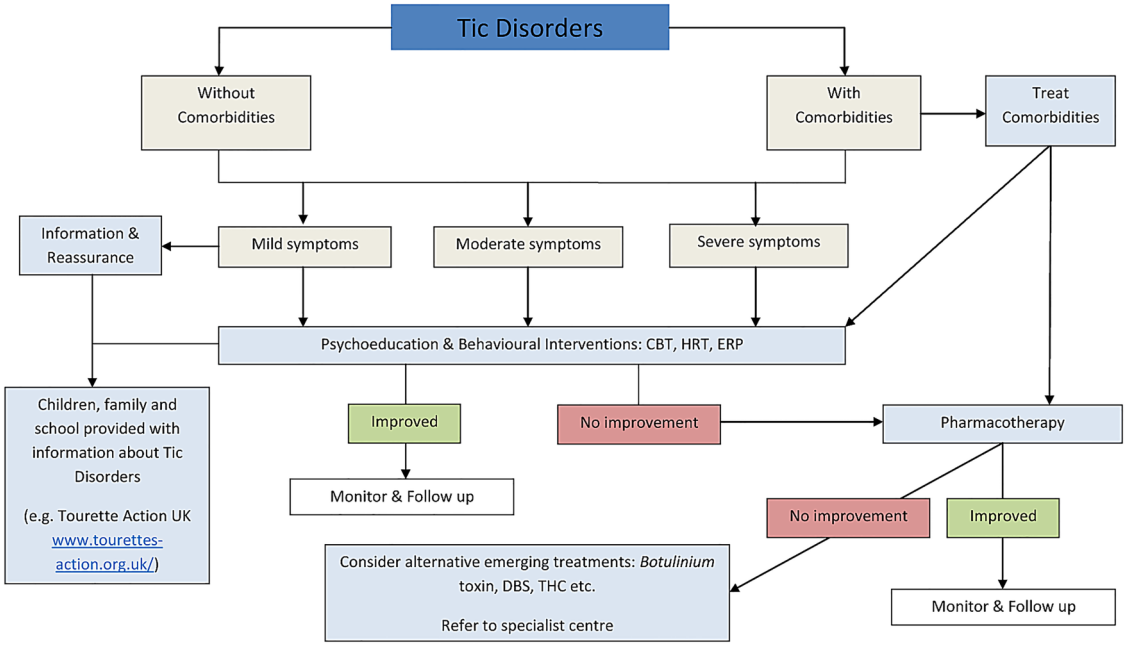

Figure 3 Suggested management algorithm for children with tic disorders. CBT, cognitive-behaviour therapy; DBS, deep brain stimulation; ERP, exposure with response prevention; HRT, habit reversal training; THC, tetrahydrocannabinol.

Table 7 Indications, efficacy and monitoring requirements of medications used for treatment of chronic tic disorders and TS

Medication	Indication	Investigations and monitoring	Frequent adverse reactions	Level of evidence	Dose
Alpha adrenergic agonists					
Clonidine	ADHD/TS	BP, ECG	Orthostatic hypotension, sedation	A	0.05 mg/day up to maximum 0.3 mg/day ÷ three times a day
Guanfacine	ADHD/TS	BP, ECG	Orthostatic hypotension, sedation	A	0.5 mg/day up to 4 mg/day
Noradrenaline reuptake inhibitor					
Atomoxetine	ADHD/TS	BP, ECG, transaminases	Nausea, dry mouth, anorexia, insomnia, fatigue, headache	A	40–60 mg/day
Typical neuroleptics					
Haloperidol	TS	Blood count, ECG, weight, transaminases, neurologic status, prolactin	EPS, sedation, increased appetite	A	0.25 mg/day up to maximum 6 mg/day ÷ three times a day
Pimozide	TS	Blood count, ECG, weight, transaminases, neurologic status, prolactin	EPS, sedation, increased appetite	A	0.5 mg/day up to maximum 10 mg/day
Atypical neuroleptics					
Risperidone	TS/DBD	Blood count, BP, ECG, weight, electrolytes, transaminases, prolactin, blood lipids, glucose	EPS, sedation, increased appetite, orthostatic hypotension	A	0.5 mg/day up to maximum of 6 mg/day
Olanzepine	TS/OCB	Blood count, BP, ECG, weight, electrolytes, transaminases, prolactin, blood lipids, glucose	Sedation, increased appetite, akathisia	B	5–10 mg /day
Benzamides					
Sulpiride	TS/OCB	Blood count, ECG, weight, transaminases, prolactin, electrolytes	Problems with sleep, agitation, increased appetite	B	100 mg/day up to 800 mg/day ÷ twice a day

Evidence level: A (more than two controlled randomised trials), B (one controlled, randomised trial), C (case studies, open trials).
ADHD, attention-deficit hyperactivity disorder; BP, blood pressure; DBD, disruptive behaviour disorder; EPS, extrapyramidal symptoms; OCB, obsessive-compulsive behaviour; TS, Tourette syndrome.

ALTERNATIVE AND EMERGING THERAPIES

It is recommended that in severe cases of TS not responding to conventional therapies other agents could be tried in specialised centres[19] (see algorithm). This includes the use of tetrahydrocannabinol, botulinum toxin injection, deep brain stimulation and transcranial magnetic stimulation.

SCHOOLS AND TICS

Providing educators with general information relating to the aetiology, presentation and course of TS will help them to implement effective individualised strategies to manage classroom behaviour, thus maximising child's learning potential.[20] There is good resource on Tourette Action UK website for educators (http://www.tourettes-action.org.uk/?&filter=teachers).

CONCLUSION

Tic disorders are common and are increasingly referred to paediatricians from primary care physicians, therapists and from local education authorities. A better understanding and support is needed for children and families. If these children are adequately managed with support from colleagues in psychology and educational authorities, they can achieve their maximum potential.

Test your knowledge

1. Jake (8 years old), a fit and well boy has had grunting and eye-blinking tics for the last nine months for most days without a tic-free period of >2 weeks. What is his current diagnosis?
   A. Provisional/transient tic disorder
   B. Chronic motor tic disorder
   C. Chronic motor and vocal tic disorder
   D. Tourette syndrome
2. Which two comorbid conditions are most common with Tourette syndrome?
   A. Conduct disorder and ADHD
   B. Depression and OCD
   C. ADHD and OCD
   D. ADHD and specific learning difficulties
3. What should the initial management be of a 9-year-old girl with mild obsessive compulsive behaviour, shoulder shrugging and simple phonic tics for more than a year who is doing well at school and has good social support?
   A. Psycho-education
   B. Habit reversal therapy
   C. Manage her obsessive compulsive behaviour
   D. Atypical neuroleptics

Answers to the quiz are at the end of the references.

Competing interests None declared.

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Data sharing statement Data regarding personal experience are available from SRM.

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Answers

1. A
2. C
3. A

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Notes