

# Pervasive development disorder

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# Definition

- The diagnostic category of pervasive developmental disorders (PDD) refers to a group of disorders characterized by delays in the **development** of **socialization** and **communication** skills.

Symptoms may include problems

- ✓ with using and understanding language;
- ✓ difficulty relating to people, objects, and events;
- ✓ unusual play with toys and other objects;
- ✓ difficulty with changes in routine or familiar surroundings, and repetitive body movements or behavior patterns.

Autism (a developmental brain disorder characterized by impaired social interaction and communication skills, and a limited range of activities and interests) is the most characteristic and best studied PDD.

Other types of PDD include:-

- Asperger's Syndrome,
- Childhood Disintegrative Disorder,
- Rett's Syndrome.

- The phrase 'autistic spectrum disorders' (ASD) is widely used to describe all of the conditions above, but recognizes that the disorders may overlap to some extent and that within any category there is a wide variation in severity of symptoms
- It takes into account the fact that a child with a PDD may have an IQ that is subnormal, normal, or above normal, and that family members of those with autism may show autistic-like traits in their behaviour



- Children with PDD vary widely in;-
  - abilities,
  - intelligence,
  - behaviors.
- Some children do not speak at all, others speak in limited phrases or conversations, and some have relatively normal language development.
- Repetitive play skills and limited social skills are generally evident.
- Unusual responses to sensory information, such as loud noises and lights, are also common.

## Autism or autistic disorder

### Onset

- Onset is before the first 3 years of life.
- 70% of cases do not have normal development.
- 30% of cases have clear 'setback' in the second or third year of life.

## Epidemiology

- The prevalence is between 7-28/10,000.
- The M:F ratio is 4:1, accounting for 25% to 60% of all autistic disorders.
- There is no clear association with socio-economic status.

## One study found

- predominance of boys over girls in autism.
- 60% of the autistic children are diagnosed before the age of 3 years.



## Etiology

Autism has various causes.

- Genetic causes:
  - ✓ Heritability is over 90% with higher concordance in monozygotic twins.
  - ✓ The recurrence rate in siblings is roughly 3% for narrowly defined autism, but is about 10–20% for milder variant of autism. Chromosome 2q and 7q are suspected locus.

- Environmental causes:
  - ✓ There is a moderately increase in rate of perinatal injury.
- Mnemonic RFT+PKU: (Rubella, Fragile X syndrome, Tuberous sclerosis, PKU: Phenylketonuria)
- CMV, neurofibromatosis and infantile spasms are also recognized causes.

# Signs of autism

- Impairment in making eye contact but able to do so when asked
- Lack of reciprocity and having difficulty identifying the mental state of other people
- Attachment to odd objects or non-soft objects.
- Rigid and resistant to change in routine and lack of imaginative play
- Lack of creativity and fantasy in thoughts

- Echolalia, palilalia (repeating the same phrase at the end of a sentence)
- Pronominal (pronoun) reversal
- Lack of social usage of language.
- Self injury (e.g wrist biting) and stereotyped behavior (e.g hand-flapped, nodding, rocking)



# Classification

diagnostic criteria include:

- The presence of abnormal development is manifested before the age of 3 years: including abnormal receptive or expressive language, abnormal selective or reciprocal social interaction, and abnormal functional or symbolic play.
- Abnormal reciprocal social interactions include failure in eye gaze and body language, failure in the development of peer relationships, lack of socio-emotional reciprocity, and lack of spontaneous sharing with other people.
- Abnormal communication includes a lack of development of spoken language, lack of social imitative play, failure to initiate or sustain conversational interchange, and stereotyped and repetitive use of language.



## Cont'd.....

- Restricted, stereotyped, and repetitive behavior include preoccupation with stereotyped interest, compulsive adherence to rituals, motor mannerisms, and preoccupation with part-objects or nonfunctional elements of play materials.
- Other non-specific problems include: phobias, sleeping and eating disturbances, temper tantrums, and self-directed aggression.
- The absence of other pervasive development disorders, socio-emotional problems, intellectual disability, and schizophrenia-like symptoms.

# Autism spectrum disorder

This is now considered a new DSM-5 terminology that now comprises 4 previous DSM-IV-TR conditions;-

- autistic disorder,
- Asperger's disorder,
- childhood disintegrative disorder and
- pervasive developmental disorder not otherwise specified

# Autism Spectrum Disorder

- The DSM-5 Criteria states that an individual diagnosed with Autism spectrum disorder would have marked difficulties in terms of communication and engagement with others across multiple social situations.

These difficulties include:

- a) difficulties with demonstrating appropriate behaviors in social contexts,
- b) difficulties associated with nonverbal communication used for social interactions and
- c) difficulties associated with failure to initiate or adapt to social interactions.

- In addition, the DSM-5 also requires individuals to have characteristic repetitive behavioral patterns, such as
  - a) Repeated stereotypical movements or
  - b) Highly ritualized behavioral patterns.
- DSM-5 criteria specified that these behaviors must have started since the early developmental period and have resulted in marked impairments in terms of functioning.



# DSM-IV diagnostic criteria for autistic disorder

A

1. Qualitative impairment in social interaction as manifested by at least two of the following:
  - marked impairment in the use of multiple nonverbal behaviors;
  - failure to develop peer relationships appropriate to developmental level;
  - a lack of spontaneous seeking to share enjoyment, interests, or achievements with others;
  - lack of social or emotional reciprocity.



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2. Qualitative impairments in communication as manifested by at least one of the following:

- delay in, or total lack of, the development of spoken language;
- in individuals with adequate speech, marked impairment of the ability to sustain a conversation with others;
- stereotyped and repetitive use of language or idiosyncratic language;
- lack of varied, spontaneous make-believe play or social imitative play.

3. Restricted repetitive and stereotyped patterns of behavior, interests, and activities, as manifested by at least two of the following:
- encompassing preoccupation with one or more stereotyped and restricted patterns of interest that is abnormal either in intensity or focus;
  - apparently inflexible adherence to specific, nonfunctional routines or rituals;
  - stereotyped and repetitive motor mannerisms;
  - persistent preoccupation with parts of objects.

B

- Delays or abnormal functioning in at least one of the following areas, with onset prior to age 3 years:
  - social interaction,
  - language as used in social communication, or
  - symbolic or imaginative play.

C

- The disturbance is not better accounted for by Rett's disorder or childhood disintegrative disorder

# assessment

Evaluation includes:

- a) An autism-specific developmental history
- b) Direct observations
- c) Obtaining wider contextual and functional information

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## Instruments

- 1) Autism Diagnostic Observation Schedule (ADOS) for patients and Autism Diagnostic Interview (ADI) for parents.
- 2) Child Behaviour Checklist (CBCL): High scores in: withdrawn/depressed, social problems, and thought problems syndromes significantly discriminates autism children from other disorders
- 3) IQ test: performance IQ is better than verbal IQ.

## Investigations

recommend that:

- 1) Children with ASD with the following features should have a genetic evaluation:
  - a) Microcephaly or macrocephaly
  - b) A positive family history (of a genetic syndrome)
  - c) Dysmorphic features.

2. Electro-encephalography (EEG) is not routinely recommended in children with ASD but should be considered if any of the following are present:
- a) Clinical seizures
  - b) Symptoms suggestive of sub-clinical seizures such as staring spells
  - c) A history of developmental regression.

## Management

For the child:

- Every preschool child diagnosed with autism should have an individualized intervention plan that sets out the goals, type(s), frequency and intensity of intervention, in order to address particular developmental and educational needs.
- An individualized intervention plan should consist of a variety of quality programmes and activities.
- This includes attendance in comprehensive early intervention programmes, programmes targeting specific needs and also positive engagement with parents and/or caregivers.

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- Alternative-augmentative communication systems may be recommended for preschool children with autism because expanded (spoken or written) communication may stimulate speech acquisition in non-verbal children and enhance expression in verbal children.
- Visual strategies are useful interventions for children with autism because they offer visual support to communication, and increase spontaneous imitation and socially communicative behavior.



- Social skills programmes depend on the functioning level of the preschool child with autism and may include:
  - Assessment and teaching of social skills interaction in natural settings.
  - Provision of structure, visual cues, and predictability.
  - Making abstract concepts more "concrete".
  - Activities that enable purposeful and appropriate interaction with typically developing peers.
  - Goals focusing on fostering self-appreciation and self-esteem.

## Pharmacological treatments:

- Fluvoxamine may be considered for repetitive thought and maladaptive behavior but should be used with caution in children with autism because of limited efficacy and poor tolerance.
- Risperidone is recommended for the management of irritability, hyperactivity and stereotypic behaviour when used as short term treatment for children with autism.
- Methylphenidate may be considered for treating hyperactivity in children with autism, although the magnitude of response is often less than that seen in typically developing children with attention deficit hyperactivity disorder.

## For the parents

- 1) Parents and caregivers should be encouraged to discuss the need for practical emotional support. This enables information to be provided, referrals made and support services made available
- 2) Parents and caregivers are recommended to consult appropriate professionals when considering educational placement for their child with autism, e.g. child and educational psychologists who are informed of the special educational provisions

## Comorbidity:

- ✓ Generalised intellectual disability (50% have  $IQ < 50$ ; 70% have  $IQ < 70$ ; 100% have  $IQ < 100$ ),
- ✓ Seizure (25%): peak age of seizure is 11-14-year-old.
- ✓ OCD (10%).
- ✓ Hyperactivity,
- ✓ Emotional problems,
- ✓ Temper tantrums.



## Differential diagnosis

- Asperger's syndrome
- Deafness. This can be excluded by appropriate tests of hearing.
- Specific developmental language disorder, in which the child usually responds normally to people, but has deficits in speech and language.
- Learning disability, in which there is general intellectual retardation with relatively less language impairment than in autism and a more normal response to other people.



# Prognosis

- The most important predictor is childhood IQ and presence of speech by 5-years.
- 50% do not develop useful speech.
- Non-verbal IQ < 60 is associated with severe social impairment and lack of independent living.
- Only 10% are able to work independently.

# Asperger's syndrome

- This is a less severe form of PDD with later onset, normal intelligence and language development and schizoid personality.
- Pedantic speech and a preoccupation with obscure facts often occur.
- Epidemiology:
- 3-4 per 1000 children; M:F = 9: 1

ICD-10 diagnostic criteria:

- 1) Relatively normal early development and the child is noted to have a lack of warmth and interest in social relationships around the third year of life.
- 2) Language development is not delayed and single words should have developed by age of 2 and communicate phrases by age of 3. Motor milestones are delayed

Asperger's syndrome is a condition in which a child has the characteristic impairments in social interaction and repetitive behaviors or restricted interests that are seen in autism, but has normal speech and intellectual abilities.

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The typical clinical presentation is of a child with the following characteristics:

- developmentally appropriate speech and language;
- unusual use of language (e.g. abnormal rate/ rhythm/volume, novel words);
- motor clumsiness;
- unusually deep interest in one particular topic;



cont'd

- rigid behaviour and stereotypes;
- social awkwardness or an eccentric social style; the child fails to read the other person's feelings, and often does not understand humor or irony;
- difficulty in making close friends;
- the child often shows an enhanced ability to rote learn information about their special interests, or perform at a high level in one particular activity

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## Comorbidity:

- Anxiety,
- depression,
- OCD,
- Possible development of schizophrenia (uncommon).

## IQ test:

verbal IQ is better than performance IQ (reversal of autism).



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## Treatment:

- 1) Psycho-education should be offered to parents to enhance acceptance and maintain routines at home and school.
- 2) As the child gets older, he can be helped by supportive counselling.
- 3) The patient is encouraged to obtain employment in routine job (e.g. librarian). Shelter employment and residence are reserved for severe cases.

## Prognosis:

- IQ>70 indicates for good prognosis. Most individuals will be able to obtain employment in a fairly routine job.
- Successful relationship with the opposite sex leading to marriage is uncommon.

# Childhood disintegrative disorder (disintegrative psychosis)

- This is very rare; the prevalence is around 2/100 000.
- It is characterized by normal initial development (to age 4 years) and then the subsequent onset of dementia with social, language, and motor regression with prominent stereotypes.
- The etiology includes infections (especially subacute sclerosing panencephalitis) and neurometabolic disorders.



# Rett syndrome

- is a rare genetic neurological **disorder** that occurs almost exclusively in girls and leads to severe impairments, affecting nearly every aspect of the child's life and their ability to;-
  - speak,
  - walk,
  - eat, and
  - even breathe easily.
- The hallmark of **Rett syndrome** is near constant repetitive hand movements

# Symptoms

- The age when symptoms appear varies, but most children with Rett syndrome seem to grow normally for the first 6 months before any signs of the disorder are obvious.
- The most common changes usually show up when babies are between 12 and 18 months, and they can be sudden or progress slowly.

## Symptoms of Rett syndrome:

- **Slowed growth.** Develops microcephaly. This stunted growth becomes clearer as the child gets older.
- **Problems with hand movements.** Most children with Rett syndrome lose the use of their hands. They tend to wring or rub their hands together.
- **No language skills.** Between ages 1 to 4, social and language skills start to decline. They may stay away from or not be interested in other people, toys, and their surroundings.
- **Problems with muscles and coordination.** This can make walking awkward.

- Children with Rett syndrome also tend to become tense and irritable as they get older.
- They may cry or scream for long periods of time, or have long fits of laughter.
- Symptoms of Rett syndrome usually don't improve over time.
- It's a lifelong condition.
- Often, the symptoms worsen very slowly, or don't change.
- It's rare for people with Rett syndrome to be able to live independently.

## Atypical autism

Atypical autism (PDD-NOS) is a diagnostic category for children who do not quite meet the diagnostic criteria for a specific PDD, but show many of the characteristics typical of them.



The End

Thank you

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