# Challenges in Anaesthesia of Children with Autistic Spectrum Disorders

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Abstract: Autism is a disease of the brain. It affects 6 in every 1000 children from all races, ethnical and social groups. Most of the physicians say it is more of a condition, rather than a disease. Children who suffer from autism have problems with communication and lack social skills. This is why they have to be under anesthesia when various medical manipulations or operations are performed. Prior to all those interventions are necessary not only consultations with pediatrician, anesthesiologists and other specialists but also close collaboration with psychologists who know the specific character of this condition.

Keywords: autism, preoperative preparation, anesthesia

#### 1. Introduction

Autism is a most general brain disorder. It affects 6 in every 1000 children from all races, ethnical and social groups. With the exception of syndrome of Rett, all its types are encountered 3 to 4 times more frequently with boys rather than with girls. Most doctors define it as a condition rather than a disease. Autistic children have problems with communication and social relations. They can have as well atypical: manner of behavior, interests and movements.

Autism is a comparatively rare disorder but in the last years globally there has been reported a statistical increase in the autistic disorder cases. During the last twenty years in Bulgaria the diagnose set-up passes from interference phase (delay in the nerve and psychic development) to the proper diagnosis. Despite this achievement there is still more to be done in direction of the early diagnostics. Frequently parents have stress by the fact that their child does not comply with the regularities pertaining to the development of other children of same age and they look for information from casual sources [1]. They are confused, anxious, feeling guilt and having many questions and few answers.

## 2. Etiology

For syndrome of Rett is typical that it appears as a result of genetic mutation. For the other types of autism reasons remain unclear despite the numerous studies. Some scientists consider that infection or environmental toxin are reasons for this disease. Others accept that autism could be displayed at brain trauma or brain disorder during the intrauterine development or during the early childhood.

Third group of researchers considers that this disease is a result of atypical levels of chemical compounds (neurotransmitters) like dopamine and serotonin that transfer the nerve signals between the cells [2].

In International Disease Codes X are described the following types of autism, and three of them are the most frequent: *1) Classic autism;* 

- 2) Syndrome of Asperger lighter type of disease;
- 3) PDD- (Not Otherwise Specified) –used to describe the group of children who do not fit in the other types of autism.
- Two types of this disease are encountered less frequently:
- Syndrome of Rett neurological disorder that appears in girls. It includes problems with movement and speech together with the remaining symptoms typical for autism.
- 5) *Child's disintegrating disorder*. This is the most severe type of this disease when child looses more physical, linguistic and social skills than in the other types.

Interpretation of autism as a generalized developmental disorder and a trinity of social, behavioral and communicative disorders in DSM III is preserved in DSM IV but in another vision – the generalized developmental disorders are being considered as particular conditions [3].

- Syndrome of Asperger deterioration of social interaction, routine and repetitive behavioral models and activity. There is no development of personal self-improvement habits corresponding to certain age, no adaptive behavior and the specific curiosity of childhood; there is no spontaneous sharing of pleasures. In the same time the manifestation of stubborn interest in parts and details of certain objects is typical. Formation of basic movements comes later, motion affectation and adherence to ritual activities are displayed.
- *Syndrome of Rett* /mainly in children of female sex/ a characteristic feature is the delayed development of physical parameter finger growth between 5 and 48 months. Mastered purposeful hand habits are lost or there appear strange hand movements reminding handwashing and twisting of fingers. Dynamic coordination is disturbed. During the pre-school period, difficulties in social functioning are observed.

• Atypical autism /designated as unspecific generalized development disorder/ - it can be diagnosed when the child's activities are less than the activities in syndrome of

L.Kanner. Table 1 shows in comparative aspect the different symptoms between syndrome of L. Kanner and syndrome of Asperger. [Table 1]

	SYNDROME OF ASPERGER
<ul> <li>Manifests itself exclusively with boys.</li> </ul>	<ul> <li>Manifests itself with boys as wells as with girls</li> </ul>
<ul> <li>Manifests itself during the first year of life.</li> </ul>	Manifests itself during the second even the third year of
<ul> <li>Walking develops earlier than speech.</li> </ul>	life.
• There is no visual contact, others are not noticed.	<ul> <li>Speech appears before walking.</li> </ul>
• Speech does not perform communicative functions.	<ul> <li>Visual contact is fugitive, shallow</li> </ul>
<ul> <li>Problems in cognitive sphere.</li> </ul>	
Limited intellectual activities.	<ul> <li>Speech is with communicative functions.</li> </ul>
	<ul> <li>Problems in emotional sphere.</li> </ul>
<ul> <li>Restless behavior, frequently accompanied by</li> </ul>	<ul> <li>Intellectual development is within the limits of norm.</li> </ul>
extreme affect reactions.	Strange behavior.
• When hospitalized there is indifference towards the	<ul> <li>When hospitalized there is desire to return back home,</li> </ul>
new setting.	but also indifference to the missing relatives.

Table 1: Comparison between syndrome of L.Kanner and syndrome of Asperger [4]

In literature were defined the basic manifestations of autistic disorder [3]:

- a) Qualitative disorders in social integration noticed in:
- missing or limited abilities of inter-personal perception;
- missing or abnormal requirements for comfort at distress situation;
- missing or disturbed imitation;
- missing or abnormal social game;
- considerable disturbances in the abilities to build up friendly relations
- b) Qualitative disorders in verbal and non-verbal communication as well as in creative work:
- There are no communication instruments like facial expression, mimics, gesticulation or in spoken language;
- There are abnormal models for non-verbal communication not using eye contact or body position;
- Lack of creative activities such as playing the role of adults, image-fantasies or images of animals, lack of interest towards fairy tales;
- Disturbances in speech production, such as content, rhythm, intonation, strength and pitch;
- Speech forms and content are disturbed;
- Disturbed abilities to begin or maintain conversation.
- c) Presence of repeating activities and interests demonstrated in:
- Stereotypical body movements such as complex body movements, monotonous shaking of one's head;
- Persistent interest in parts of different objects;

- Distress demonstration when trivial aspects of surrounding media have been changed;
- Misunderstanding of changeable punctually defined details and change;
- Interest only in one way.

Autistic children demonstrate no or slight interest towards the others including towards the members of their family [5]. There is no motivation for social integration. The deficit in linguistic and speech development appears to be a clear indicator. Most of the autistic children are non-verbal. They either establish elementary communicative acts with prevailing non-verbal and scanty usage of signs or demonstrate echolalia, perceiverations or usage of words that are not included in the standards of any language [4].

Definitive for the differential diagnostics is the so called trinity of social interaction: disturbance of social interrelations, disturbance of social communication and disturbance of social understanding and presentation [4], [7].

Specific monitoring features could be: lack of any selfdefense reactions, fear (from height, animals, darkness ect.), unspecific reactions of pain ( its location is not pointed out, pain is not expressed in habitual way, its intensity is not well defined etc). Due to the behavioral disorders autistic children react in atypical mode to stomachache, and frequently it is not "decoded" by their parents [8]. In general, Table 2 presents five cases from personal practices, which have been consulted in the period 2010 - 2015. [Table 2]

Table 2:	Cases from	personal pr	actices, w	hich have b	been consulted	in the	period 2010 – 2	2015
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Group	Indicators	I.M.V. –boy, 5	E.T.P girl, 4	A.N.N. –boy,	A.S.M. –girl, 5	S.T.Sboy, 3 yrs.
of		yrs. 3 m.	yrs.	6 yrs. 7 m.	yrs.	5 m.
indicators						
tie	Disability to use eye	Lack of visual	Lack of visual	Lack of visual	Slight visual	Lack of visual
ilal	contact, body position,	contact,stereo	contact. Hyper	con tact, stereo	contact. Changes	contact. Not well
n soc	facial expres sion and	type body	active. Not well	typic hand	her expression	coordinated when
ctic al	gestu res for adequate	movements. Does	coordina	movements. In	spontaneously.	walking. Does not
ab roc	interaction regulation.	not use gestures to	ted. Destruc tive	some moments	Impulsive.	feed himself.
ive cip. nte		state a certain	towards material	is chaotic.		
rec		need.	objects.			
ali in	Lack of development	Lack of cognitive	Specificity of	Can be	Moderate	Does not interacts
	corresponding to mental	interest Attention	development	monitored	mentally	with children of the

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age and despite of the existing possibilities, of relations with children of the same age, inclu- ding common interests, attitu-des and emoti- ons,		is of no deliberate nature. Neces-sity of comm unication with others is not monitored.	Recognizes colors, has some concepts already shaped. Sporadically establishes contact with other adult. There is a need of stimulus material.	After mother's data, the boy can play pia- no. Has a specific me-mory for movements' sequence.	retarded. Well influenced by specialized complex care. Interacts with other children.	same age or adults.
	Rarely looks for or uses other people.	Does not seek his parents and other people.	Does not seek her parents and other people.	Partially. Attached to his mother.	Has emotional contact with the mother.	Does not seek his parents and other people.
	Lack of sharing	It is present. Non- verbal.	It is present.	It is present.	It is present.	It is present.
	Lack of social- emotional reciprocity.	It is present. Emotional deficit. Not attached.	It is present. Emotional deficit. Slight emotional attachment.	It is present.	Partially. Inclined to aggression and autoagression.	It is present.
	Retarded or total lack of speech which is not compensated by the usage of gestures or mimics.	Lack of linguistic structures.	Does use the mastered words(from the spoken language) to declare a need and for a contact.	Falls behind in the lingu-istic deve-lopment. Does not use gestures or mimics.	Speech is scanty, strongly restricted. Notdeliberate. Rarely it is a situational one.	Falls behind is the linguistic development.
cation	Relative difficulty to begin or maintain a conversation.	Expressed difficulty.	It is present.	It is present.	It is present.	It is present.
ities in communic	Stereotypic and repeated usage of language and/ or idiopathic use of words and phrases.	It is not monitored.	Synthetic, idiopathic, emotionally saturated "language". Not determined	Sometimes.	It is monitored. Echolalolic speech.	Stereotypic repetitions of phrases without any content.
Qualitative abnorm	Abnormality in the pitch, stress, pace, rhythm and intonation of the voice.	Not monitored.	Clear diction, good intonation. She has musical ear. Infringements in the pace.	When speaking a change in rhythm can be noticed periodically. Especially in the chaotic	It is monitored.	Not monitored.
	Lack of spontaneous game and socially imitative game.	It is present. Manipulation with toys is not considered as a game act.	Impulsive displays of a desire for dynamic game, freque-ntly she is not cooperative.	Partially.	Sometimes there is a desire for game interaction with the others.	It is present.
oehavior,	Overwhelming meditation in stereotypic and restricted interests.	Partially. Attracted by centrifugation in washing machine.	It is present.	Partially.	Partially.	When aligning objects on the table he shows pedantry.
ts of t	Specific attachment to unusual objects.	It is not monitored.	It is not monitored.	To cosmetic catalogues.	To a toy.	To clothes-pins.
cotyped print nd activities	Noticeable attachment/clinging/ to specific non-functional rituals.	It is being registered-sand, water, foam.	It is not monitored.	To opening and closing of door, cabinet, wardrobe. Cables.	To food. External control is needed daily.	Unfastening of press studs, magnets.
repetitive, ste interests a	Stereotypic and repeated motor mannerisms	They are monitored during most of the time.	They are monitored periodically.	They are monitored. All the time the mother stands next to him.	Sometimes. She turns in circle.	When an external person comes in, he immediately glues in a corner.
Restricted,	Meditation in sections or nonfunctional characteristics of the objects.	When interacting with sand, water, foam.	It is present. Not monitored all the time.	Sometimes.	Slightly expressed.	Arranging of objects.
	Distrass when tiny	It is monitored	Slight pagative	A ggravates the	It is monitored	He gets upset when

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non-funct-ional details in the media are changed.	Becomes worried, becomes affected quickly.	emotional transitions.	chaotic, behavioral models. He has a good memory for locations	Strong emotional reaction when the setting is being changed.	there are changes in the setting.
			and routes.		

Along with the fast increase of cases from autistic spectrum diseases (ASD) and the extension of debate for best treatment and therapy, the consequences for family life from a ASD diagnosis remain comparatively unstudied. In many cases the opinions of parents and professionals do not always coin-cide [9].

Anaesthesia and ASD: At early childhood age children with autism frequently need several unpleasant medical tests or procedures to be performed and this imposes the implementation of sedation or anaesthesia. This poses in front of anaesthesiologist several challenges due to the specific nature of this disease.

**Pre-operative preparation**: Most frequently the problems connected with pre-operative preparation of autistic children are bind with:

- 1) Difficulties to establish contact with them and impossibility for them to describe their complains.
- 2) Lack of desire to establish physical contact.
- They are uncooperative when some functional tests are done (nuclear magnetic resonance/NMR/, functional lung tests etc.)
- 4) Frequently they feel fear when enter unfamiliar surrounding.

The nature of this disease suggests difficult communication with the children. Frequently they have heightened anxiety, wary attitude, and sometimes they are aggressive. That is why the aneasthesiologist who should implement the anaesthesia has to apply different than the usual methods in order to gather information for these children.

This is done through a preliminary conversation with the parents (the child's guardian) about the child's physical and psychological needs. Rainey L(1998) recommends the preoperative preparation for anaesthesia to include obligatory a talk with parents dealing with the work schedule of the anaesthesia and the possible options for its implementation[10]. Sometimes for the child's comfort it is necessary to bring in the hospital their favourite toys that amuse and calm them. It is obligatory autistic children to be enlisted first in the operative schedule thus the effect of waiting and hunger will be diminished; while with the ones having cognitive deficit is recommended the parents' presence. For the ASD children is typical that more frequently they tend to oppose to the anaesthesia when in

their anamnesis they have similar opposition also towards other kind of manipulations, e.g. immunizations etc. [11]. That is why the usage of symbols familiar to them is very useful in pre-operative preparation; in other words communication is facilitated and their anxiety is considerably reduced. For this purpose with these children are implemented the so called **widgit symbols** (Figure 1, Figure 2) [12]. The pictures' sequence follows the sequence of events before, during and after anaesthesia.



Figure 1: Widgit Symbols





With children having limited speech the anaesthesiologistreanimator could make use of the sign language applying the communication system through pictures: (Figure 3) [13].



The permanent venous catheter is fixed like with the other children. The preliminary skin treatment with EMLA at the puncture point or induction with inhaling anaesthetics are proper techniques in such cases.

**Premedication**: Per-oral premedication is preferred [14], [15], [Table 3]. Midazolam application in doses higher than 0,5 mg/kg leads more frequently to paradoxical dysphoria. Despite this its fast beginning and short time effect, as well as the lack of side effects make it an attractive medicine in these cases [16].

Per-oral Midazolam application is considered as more effective with children having lighter degree of autism. It can be diluted in the child's favourite juice. Ketamin is another medicine that due to its sedative and analgetic qualities also finds a wide application with these children [17].

Preferred per-oral dosage - 8 mg/kg.

Some authors consider its independent application in premedication as not so effective.[16](Ghai B.). It is used in moderate to severe cases of autism. Nevertheless, the combination of both medicines is preferred by most authors, in other words they neutralize mutually to a certain extend the side effects[14], [18], [19], [20].

In several studies, the successful application of alphaadrenergetic agonists is also being proved.

Per-oral application of Klonidin at dosage 2-4 mcg/kg has excellent sedative and anxiolytic effect with ASD children[14], [21]. In 1990 Dexmedetomidine was presented; it is another medicine that became popular in more than 1100 published reports dealing with its application. It is attractive for premedication, i.e. it has reliable sedative effect, it has excellent safe profile with minimal risk of defying respiratory depression and changes in haemodymanics [22].

Its per-oral application for anaesthesia induction or for sedation is effective even with patients with behavioral disorders who have had previous, unsuccessful attempts for sedation [23].

Nevertheless, due to the considerable suppression of sinus and atrioventricular nodal function with children, it is not suitable with these having bradycardia or antrioventricular block [23].

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Medicine	Manner of application	Dosage	Maximal dosage	Start of action
Midazolam	РО	0.25-1 mg kg <sup>-1</sup>	20 mg	20-30 min
Ketamin	PO	3-5 mg kg <sup>-1</sup>		30-60 min
	IM	1-2 mg kg <sup>-1</sup>		~ 3 min
Midazolam +	PO	0,5 mg kg <sup>-1</sup>	20 mg	~ 30 min
Ketamin		3 mg kg <sup>-1</sup>		
Klonidin	PO	4 mkg kg <sup>-1</sup>	150 mkg	45-60 min
Lorazepam	PO	50-100 mkg kg <sup>-1</sup>	4 mg	~ 60 min

Table 3: Premedication in children with special needs including the ASD ones: [12].

# Anaesthesia and possible complications after anaesthesia:

Some risk factors leading to unwanted complications after anaesthesia are:

1)Anamnesis of convulsions

2)Pre-operative respiratory problems.

3)Poor clinical condition of child before the procedure.

4)Not diagnosed mitochondrial disease (disorder in energy production, mitochondrial myopathy, known also as mitochondrial oxidative phosphorylation - OXPHOS ).

All these impose during the preoperative period to be tested indicators as:

1)Complete metabolic profile;

2)Serum magnesium levels;
3)Differential complete blood test;
4)Serum creatinine-kenase;
5)Serum amylase;
6)Serum ammonium level;[24]

**Post-operative anaesthesia**: Children with special needs have to be well anaesthetized, i.e. during the post-operative period it is difficult to make difference between crying due to operative pain and due to other reasons like nausea, fear of the unfamiliar setting etc. In these cases the anaesthesiologist is assisted by the behavioral pain scales. [Table 4]

 Table 4: Behavioral pain scale

Applied to patients who can not report the pain presence without outside help: estimation from 0 to 10 at clinical

monitoring.							
Face	0	1	2	Face			
	Loose facial	Facial-muscle tension,	Frequently or most of the	Result:			
	muscles	they frown and make	time are frowned, tensed				
		grimaces	lower jaw				
Anxiety	0	1	2	Anxiety			
	Quiet, calm	Rarely affected nervous	Frequent nervous	Result :			
	vision, normal	move-ments and	movements, which can				
	movement	change in body position	include head and limbs				
Muscle tonus*	0		2	Muscle tone			
	Normal	Higher tone, flexion of	Rigid tone	Result :			
	muscle tone	hands and legs fingers	2011				
Vocalization **	0	7	2	Vocalization			
	There are NO	Casual moanings,	Frequent or continuous	<b>Result</b> :			
	unusual	shouts, moaning and	moans, cries, whimpers or				
	sounds	groaning	grunts				
Possibility to be	0	1	2	Possibility to be			
clamed down	There is	Children are calmed	It is difficult to be	calmed down			
	content, calm	when touched, can be	comforted by touching or	<b>Result :</b>			
		distracted	conversation				
Babaviaral scale for p	ain is actimated n	urguant tan grada goala (	0 10)	0/10			

Behavioral scale for pain is estimated pursuant ten–grade scale (0-10)

\* Estimation of muscle tone in patients with lesion of spinal cord or with a wound above the lesion location. It estimates patients with hemiplegia on the side that is not damaged.

\*\* This element can not be measured in patients on artificial lung ventilation.

## 4. Conclusion

Frequently the children from autistic spectrum (ASD) are in need of medical tests, manipulations or operations that impose the implementation of sedation or anaesthesia. The nature of their disease is connected with an increased anxiety level, frequently they do not respond, sometimes they can be aggressive and this imposes necessity to have extensive knowledge of this kind of conditions. Adequate pre-operative preparation is necessary in order the children with this disorder to receive successfully implemented anaesthesia.

This preparation requires not only the habitual for every surgical intervention consultations with pediatrician, anaesthesiologist and other specialists, but also close collaboration with psychologists who are well acquainted with the specific character of this condition. Unfortunately, in Bulgaria there is still no team approach in the work among anaesthesiologists-reanimators and psychologists; this poses several questions for the active role of psychologist in pre- and post-operative preparation and care for these children.

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2319