

# Management of Children with Intellectual and Developmental Disability in an African Setting

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## 1. Introduction

Disability is an umbrella term, covering impairments, activity limitations, and participation restrictions. Impairment is a problem in body function or structure; an activity limitation is a difficulty encountered by an individual in executing a task or action; while a participation restriction is a problem experienced by an individual in involvement in life situations. Thus disability is a complex phenomenon, reflecting an interaction between features of a person's body and features of the society in which he or she lives (WHO, 2011). This issue is so important that World Health Organization (WHO) and World Bank Group command a report on it (WHO, 2011). People who experience mental health conditions or intellectual impairments appear to be more disadvantaged in many settings than those who experience physical or sensory impairments. Prevalence of children with disabilities vary substantially depending on the definition and measure of disability. Available data suggests that 93 million (5.1%) children aged 0-14 years have moderate or severe disability" (WHO, 2011). This matter is poorly addressed in Africa especially in sub Sahara region. We present here childhood intellectual and developmental disability management in an African setting. We will cover in this prospective survey, diagnosis tools regarding the clinical practices (clinical examination, psychomotor evaluation) with contribution from pediatricians, specialists of psychiatrics and neurologists; laboratory investigations with insight on South-North collaboration in medical genetics sector (cytogenetics and molecular biology); main etiologies; management; social considerations and perspectives in intellectual and developmental disability management in Benin and in West African sub region.

## 2. Patient and methods

The present survey was conducted in the Pediatrics and Medical Genetics Service of the National Teaching Hospital of Cotonou in Benin. Cotonou is the main city of Benin, a low income country in West Africa located between Nigeria and Togo. Health services are still poor and the under 5 morbidity and mortality are related to malaria, pneumonia and diarrhea (Black et al., 2010). Health conditions such as birth asphyxia and congenital abnormalities exist (Black et al., 2010). Birth asphyxia and birth defects are known to cause neurodevelopmental impairment with intellectual and developmental disability (Wright et al., 2009). But some efforts were made to cover these matters with the implementation of medical genetics services in the Pediatric ward (Alao et al., 2008; Alao et al., 2010). It was the

unique place where people can receive genetics services in Benin. Medical genetics consultation was carried out by a pediatrician who specialized in clinical genetics assisted by two nurses. This consultation took place once a week and three persons were checked per session. This prospective and descriptive study was conducted from January 1st 2009 through December 31st 2010. The recruitment was systematic with the fulfillment of a questionnaire by the medical crew. Each consultation took 45 to 60 minutes with four steps that were medical history record, psychomotor evaluation, dysmorphological examination and a general physical examination. Medical history record is a very important moment because it helps collecting any information that could lead to genetic disease or environmental factor. The previous birth of a similarly affected child suggests that the condition is likely to be genetic in origin. It is, however, important to consider the possibility of the mother taking a teratogenic drug in successive pregnancies or that the mother is alcoholic, which could result in more than one child being born with fetal alcohol syndrome. Parental consanguinity may suggest that the patient had an autosomal recessive disorder. Maternal drug/alcohol intake or any other teratogenic agent must be revealed. Maternal illnesses such as viral infections in the antenatal period were run out since they could result in the birth of a baby with mental retardation (suri, 2007). We also collected data regarding delivery, neonatal period, infant period and childhood. Informations were obtained on onset age and progression, the quality of sleeping, nutrition, social behavior and status regarding being clean by day and night. This record ended by family's tree drawing (Goldenberg and Saugier-Weber, 2010). Psychomotor evaluation was based on gross motor milestone checking. We controlled if the patient had achieved on time the expected milestone according to his or her age which was expressed in months. The six milestones studied were sitting without support, standing with assistance, hands-and-knees crawling, walking with assistance, standing alone and walking alone. Concern was expressed only if the child has not performed one or more milestones that he or she should have achieve on expected time (WHO, 2006). This psychomotor evaluation were completed by primitive reflexes and hypotonia monitoring in the young infant which were not supposed to start sitting according to their age. This stage was said to be less than 4 months. Normally these primitives reflexes were known to disappear before three months and at that time infant must be able to control his or her neck. So the persistence of primitive reflexes or failing in controlling neck position was categorized in psychomotor delay. Autism spectrum was suspected in children if the onset story was before the age of three years and these three impairments were present: social interaction disability, communication disability and restricted repetitive behavior. Social interaction disability regards lack of response to other people's emotions and deficient of inappropriate use of social signals; communication disability is related to the lack of social usage of whatever language skills are present, impaired imagination, a "mechanical" style of expression, with little flexibility or variation and lack of accompanying gesture to add meaning to communication. Restricted repetitive behavior seems to an apparent preference for rigidity and routine in a wide range of aspects of daily living such as insistence to perform routines in nonfunctional rituals, motor stereotypies, stereo typed interests, resistance to change in personal environment (WHO, 1996). When communication was poor without the full spectrum, patient was considered having autistic trait. The third time of this examination was reserved to the detection of any minor or major physical abnormalities regarding dysmorphology. The principles that were used were conformed to the Jones smith book (Jones, 1997). This took into account particular parts of the body. Craniofacial abnormalities could be found at the head, face, hand and feet, neck, chest, abdomen, skin, external genitalia and spine. Head examination could disclosed microcephaly, macrocephaly, craniosynostosis with scaphocephaly, due to sagittal

synostosis, brachycephaly from premature closure of the coronal sutures or plagiocephaly from closure of only one coronal or lamboid suture and scalp defects. Face examination could lead to discover abnormalities such as facial cleft, asymmetry, coarse face, small/narrow/elongated/ broad face, round-shaped, square-shaped, triangular-shaped, flat, hypoplastic malae, midface hypoplasia, full cheeks. Hands and feet could showed single transverse palmar crease, deep palmar and plantar creases, polydactyly, syndactyly, oligodactyly, ectrodactyly, camptodactyly, arachnodactyly, absent or hypoplastic nails, and terminal transverse defects of the fingers and toes. Ulnar ray and radial ray defects could be seen in some patients. The neck findings that one should specifically be looked for include short webbed neck, torticollis, branchial pits or sinuses and thyromegaly (Suri, 2005; Mercks et al., 2003). Skin anomalies and neurological status with tonus and reflexes testing were also done. General physical examination was done to detect problems which could be found in the other systems or organs such as locomotors, digestive, cardiologic, respirator, urologic, genital and ear, nose and throat. The examination was terminated by hypotheses' evocation. Pictures were taken with patient's permission and signed consent form in which it was stated that iconography and data could be used for scientific communications and publications in the respect of strict anonymous. Laboratory investigations generally encompassed karyotype and deoxyribonucleic acid extraction when single gene disorder was suspected. These genetic investigations were carried out in Cotonou in the Cytogenetics Laboratory of the Faculty of Health Sciences (Gangbo et al., 2010). Karyotype was done on whole blood which was collected in tube with heparin sodium. If needed, molecular tests were performed abroad through scientific cooperation with some genetics centers in Europe such as Human genetics centre of Leuven, Belgium; Institut of Pathology and Genetics of Liege, Belgium; Biochemistry and molecular genetics laboratory, Hôpital Cochin, Paris, France; Functional cardiogenetics and molecular and cellular myogenetics Unit, Hôpital Pitié Salpêtrière, Paris, France and Human genetics laboratory, Université Victor Segalen, Bordeaux, France. Extract deoxyribonucleic acid was sent to them either by postmail or by a person that was travelling to the dedicated laboratory place. Deoxyribonucleic acid was extracted with phenol/chloroform method on whole blood (Adeli and Ogbona, 1990). Patients were reviewed monthly until the suspected diagnosis was confirmed (or refuted). Follow up was established for individual assistance. This included a medical component which help preventing and detecting known complications regarding each disorder. A supportive care was planned for each patient with referral to physiotherapist, psychologist and speech therapist if appropriate. Prenatal diagnosis was also proposed if it was thought to be relevant especially in chromosomal aberration situations. Data upon socio demographic and clinical patients' characteristics (referral reasons, origin of referral, psychometric performance, etiologies, external birth defects, outcome and bad prognosis factors), contributions from specialists (medical and supportive) and school attending rate were collected. There were treated and analyzed with Epi info software package version 3.5.3. Proportions were compared with chi square, Pearson's correlation test and difference was thought to be statistically valuable when  $p < 0.05$ .

### 3. Results

A total of 206 patients were received for intellectual and developmental disability during these two years of survey. They were categorized having intellectual and developmental disability since they failed to achieve the WHO labeled six gross motor development milestones. Some showed global psychomotor delay (58%) while the other seemed to be mentally retarded (42%). Six cases of autism spectrum disorders were suspected in children

(5 males and 1 female) while 25 children bore autistic trait. Most of the patients that were enrolled in this survey (81%) were outpatients and were sent by independent pediatricians. The others were seen during their hospitalization for many different purposes. Infants were mostly recruited since they accounted for 65% of the survey population. Sex-ratio was 1.39 as shown in table 1. Almost all of the patients were referred for intellectual and developmental disability. In all, 84% were received for psychomotor delay while the others were seen for a variety of facial dysmorphism with 10% (Fig. 1, 2, 3, 4, 5, 6) or birth defect with 6%. The birth defects included palate and lip clefting, acrocephalosyndactyly, hexadactyly, congenital heart abnormalities mostly atrioventricular canal defect (Fig. 7, 8). The etiologies were dominated by birth asphyxia, followed by Down syndrome (Fig. 9, 10, 11, 12, 13, 14) and neonatal jaundice as shown in the table 2. Single gene disorders diagnosis were confirmed through international collaboration network. Thus, mutations in Fibroblastic Growth Factor -Receptor 2 (c.252C> G and c.253C> G) were identified in the Center for Human Genetics of Leuven in Belgium in Apert syndromes, c.7783G> A mutation in exon 62 of Fibrillin1 gene has been identified in Marfan's disease after research in Functional cardiogenetics and molecular and cellular myogenetics Unit, Hôpital Pitié Salpêtrière, Paris, France. Di George diagnosis was made by Fluorescence hybridation in situ at the Institut of Pathology and Genetics of Liege, Belgium. Some patients were sent to other specialists either for consultation or for supportive care as presented in table 3. Patients were sent to neurologist whenever they showed symptom or behavior that indicated possible minor epileptics. These were mainly recruited in birth asphyxia group. Cardiologic consultation was required systematically for all patients that bore Down syndrome. The patients especially infant with autistic spectrum were referred to psychiatry to exclude a real and total autism. All of the patients with poor tonus and no standing status were sent to the physiotherapist while some with no or uncompleted language had to see the speech therapist. Twenty children with Down syndrome and one with Di George syndrome were advised and guided towards ordinary education program but only five in the Down syndrome group and the one in the other group succeeded in attending one. This gave us if we consider only the group of child (n=64), an education rate of 9.37%. Seven deaths occurred with respectively 4, 2, and 1 in cases of Down syndrome, birth asphyxia and unknown diagnosis as illustrated in table 4. While considering outcome, infants had a high death risk since Fischer exact p-value was 0.0412 and bearing Down syndrome exposed to death than other etiology.

	<b>Infant</b>	<b>Child</b>	<b>Adult</b>	<b>Total</b>
Male	84	30	6	120
Female	50	32	4	86
<b>Total</b>	<b>134 (65%)</b>	<b>62 (30%)</b>	<b>10 (5%)</b>	<b>206</b>

Table 1. Patients' gender and age.

	<b>Frequency</b>	<b>Percentage</b>
Birth asphyxia	61	29.6
Down syndrome	41	19.9
Neonatal jaundice	20	9.7
Other genetic causes*	8	3.9
Unknown	76	36.9
<b>Total</b>	<b>206</b>	<b>100</b>

Table 2. Etiology of intellectual and developmental disability.

\*This included one case of Patau syndrome (Fig. 15), Di George syndrome (Fig. 16), Aarskog syndrome (Fig. 17), and Marfan syndrome; two cases of Hurler syndrome (Fig. 18) and two other cases of Apert syndrome (Fig. 19).

Specialty	Sent	Gone
Neurology	36	25 (69.44%)
Cardiology	41	21(51.21%)
Psychiatry	10	6 (60%)
Physiotherapy	134	62 (46.26%)
Speech therapy	43	15 (34.88%)

Table 3. Other specialties’ usage.

Outcome	Infant	Child	Adult	Total
dead	7	0	0	7
Alive	100	53	7	160
lost	27	9	3	39
<b>Total</b>	<b>134</b>	<b>62</b>	<b>10</b>	<b>206</b>

Table 4. Outcome and age.

**Figures**



Fig. 1. Craniofacial dysmorphism with microcephaly, upslanted palpebrale fissures, strabism, large pinea, long and large philtrum and relative macrostomia.

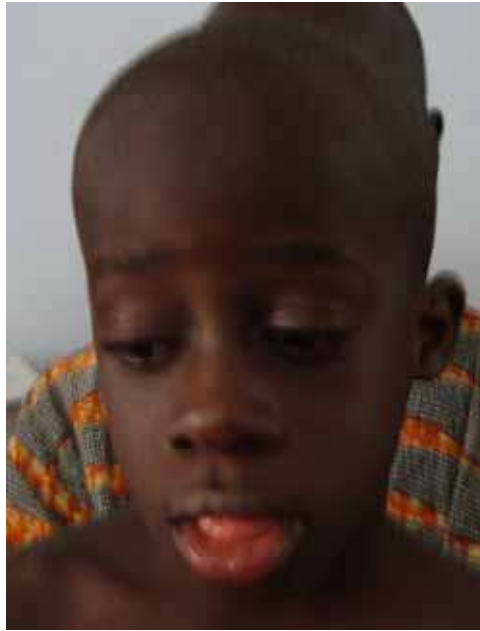


Fig. 2. Craniofacial dysmorphism with Brachycephaly, triangular face, large pinea, broad nose, relative macrostomia and lingual protuding.

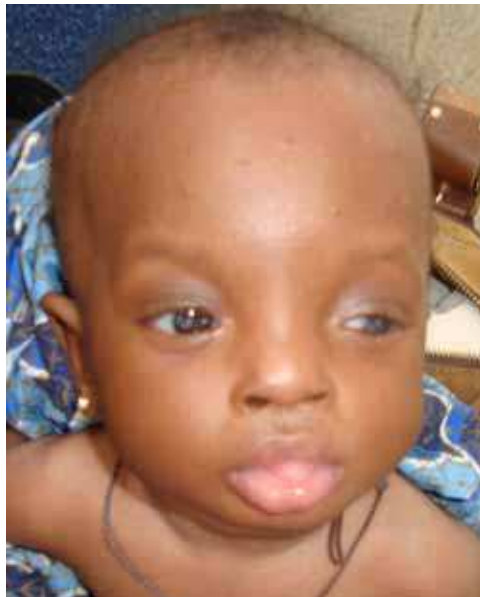


Fig. 3. Craniofacial dysmorphism with microcephaly, strabismus, broad nasal bridge, anteverted nostrils, short and large philtrum and lingual protuding.



Fig. 4. Craniofacial dysmorphism with bombing front head, hypertelorism, upturned nose, webbed cheeks, unmarked philtrum and curved linear groove below lower lip



Fig. 5. Craniofacial dysmorphism with brachycephaly, relative exophthalmos, small and tapered nose, anteverted nares, webbed cheeks and downturned mouth.



Fig. 6. Craniofacial dysmorphism with microcephaly, hypertelorism, upslanted palpebral fissures, large pinnae, and light facial asymmetry.



Fig. 7. Birth defect with lateral lip clefting



Fig. 8. Birth defect with complete syndactyly in a case of Apert syndrome





Fig. 9. Down syndrome facial feature in a 8 years old girl



Fig. 10. Down syndrome facial features in a 24 months old boy



Fig. 11. Down syndrome facial features in a 6 years old boy with exophthalmos and strabismus.



Fig. 12. Down syndrome facial features in a 5 years old girl with strabismus and early teeth renewal.

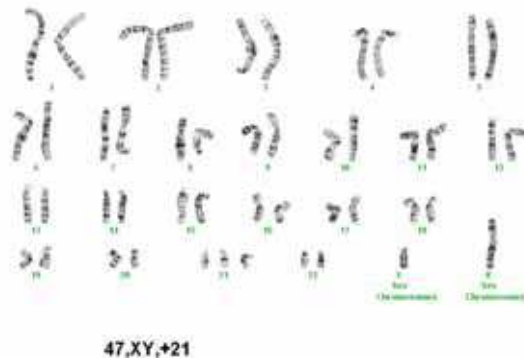


Fig. 13. Full trisomy 21 determining Down syndrome in male.

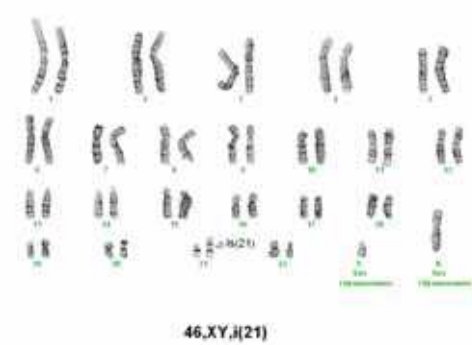


Fig. 14. Isochromosome 21 determining Down syndrome in a boy.



Fig. 15. Facial dysmorphism with coarse face, hypertelorism, big nose, relative midline dimple and single transverse palmar crease

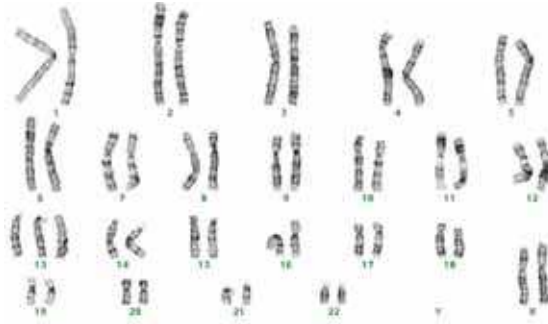


Fig. 16. Karyotype with full trisomy 13 determining Patau syndrome



Fig. 17. Facial dysmorphism in Di George syndrome with bulbous nasal tip, long philtrum and microstomia



Fig. 18. Cardiac malformation with shoe shape in a boy suffering from Di George syndrome



Fig. 19. Shawl scrotum in Aarskog syndrome



Fig. 20. Macrocephaly and coarse face with large head, bulging frontal bones, depressed nasal bridge, broad nasal tip, anteverted nostrils, full cheeks, very large ears and enlarged lips.

## 4. Discussion

### 4.1 Importance of intellectual and developmental disability

The intellectual and developmental disability cover different matter regarding the origin or the training of the person that in considered. Intellectual disabilities were referred to as illnesses, disabilities, or both, and no consensus about these terms existed. But mental

retardation is the most widely used term, although many persons also referred to intellectual disabilities. One incentive for implementation of standard use of a term that refers to disability, rather than to intellectual or mental retardation, is the fight against stigmatisation of persons with intellectual disabilities and their families (WHO, 2007). In this current survey, intellectual and developmental disability was seen as either global psychomotor delay or real and pure mental retardation. This issue has been a marginal area for health care and health research. In most countries, it receives little or no attention during medical training, and a large divide exists between availability of services and the health needs of affected individuals. A research gap is also present between intellectual disability and other neuropsychiatric disorders, which largely contributes to the invisibility of the disorder in global health policy (Shekhar Saxena, 2009). Although more than 90% of children and families affected by developmental disabilities are likely to live in developing countries, it appears that more than 90% of research, preventive efforts and services related to developmental disabilities were directed toward the populations of the world's wealthier countries (Durkin, 2002). Very few studies were reported from developing countries (Maulik et al., 2011).

#### **4.2 Prevalence of children with disabilities**

The prevalence of intellectual and developmental disabilities in children over the world varies considerably according to the level of reporting either clinical or hospital studies, urban or rural stage and taking into account pure mental retardation or global developmental delay with the lack of consensus if this matter is a disease or a disability. The real prevalence especially in developing countries will remain underreported until data collection and report are improved. According to the literature, intellectual and developmental disabilities prevalence in children could be estimated to 18.30 per 1000 populations (Maulik et al., 2011). Few reports on intellectual and developmental disabilities in Africa were available. Fifteen countries had assumed that research was done on intellectual disabilities but data were really published from few countries such as Egypt, Ethiopia, Zambia and South Africa (WHO, 2007; Maulik et al., 2011). In fact, most African countries are still fighting against communicable diseases especially in children and even if population experienced intellectual and developmental disabilities, this could not be addressed for lack of means, facilities and trained health workers (Black R et al, 2010). No study could be found on this issue in Benin as reported in the world Health Organization review (WHO, 2007). So this current report even if it is conducted at hospital level, gives some insight upon intellectual and developmental disabilities in children in Benin. This issue was poorly or not addressed in Benin because child sanitary situation is still dramatic with a lot of illiterate parents, many home deliveries, high neonatal mortality rate and infant mortality rate according to the last demographic and health survey (Institut National de la Statistique et de l'Analyse Économique, 2006). Neonatal mortality rate was at 32 per thousand live births and main causes of death were preterm birth complications, birth asphyxia, neonatal sepsis and congenital abnormalities. On the other side, infant mortality which rate was estimated to 67 per thousand live births came mainly from malaria, pneumonia and diarrhea (Black R et al., 2010). Such health care situation gives little opportunity to look at intellectual and developmental disabilities especially in children who still having low consideration in term of attention and right for decent and good life. But since genetic services became available in 2005, this issue received some attention (Alao et al., 2008). Children with intellectual and developmental disabilities were referred for consultation for research purpose because physicians and parents kept in their mind that

it's due to inheritable diseases. At that time poor consideration were given to the issue because we lack experience in evaluation and supportive care and we were oriented towards dysmorphology and birth defects regarding our back ground and training in Europe (Alao et al., 2010). Nowadays, more attention is given to intellectual and developmental disabilities in children in our consultation and the proof is in this current report. In this survey we recruited mostly children and few adults. This situation aroused from the lack of adult genetic services. It is not particular to our setting but over the world genetic services receive and manage both children and adults since infant and child become attached to the health care providers and claim to continue the follow up in the same ward.

### 4.3 Diagnosis process

Diagnosis tools that were used in this survey were compatible to what is generally recommended even if some specifics techniques were not available either in our setting or out of reach through partnership network in genetics. It is true that our diagnosis stools were mainly based on genetic practices with history record, clinical examination and laboratory investigations. The clinical examination starts by patient and family histories record. Indeed, if clinical evaluation is the key to appropriate genetic investigation in this group, then thorough history taking is the key to clinical evaluation. It is essential that the history ascertained covers as broad a time span as possible, from pregnancy onwards to capture the evolution of the presenting features. The presence of other general health problems that the parents may feel are unrelated may hold the key to diagnosis and should be carefully explored. In assessing the family history it is important to remember the concepts of penetrance and expressivity. Heritable conditions are non penetrant when individuals who carry pathogenic mutations have no signs at all of the disease normally caused by that mutation. Non penetrance is rare in neurodevelopmental disorders however variable expressivity is extremely common. It is important because it can lead doctors, and family members, to believe that a child is unaffected when in reality they are mildly affected by the same condition as their more severely affected sibling and may go on to develop complications of this at a later stage (Wright et al., 2009). Psychomotor evaluation followed the history record with variable tools. In this survey, we used World Health Organization six gross motor development milestones which was completed by primitive reflexes in young infants. This choice seemed to be suitable for our conditions. We have no other solution since we did not have more elaborated testing system with accessories and our populations were used to evaluate development by these milestones. Normally a lot of testing with intelligence quotient or development quotient determination was available but with their limits (MacLean et al., 2011). One could cite for the children the British Ability Scales, the Kaufman Assessment Battery for Children, the Planning, Attention, Simultaneous, and Successive test, the Universal Nonverbal Intelligence Test and the Wechsler Intelligence Scale for children and the Wechsler Primary and Preschool Scale of Intelligence (Sparrow et al., 2000). The Wechsler Scales are commonly most used in intellectual disability services since they seem to be simple and were administered with no difficulty. Indeed they allow the evaluation of verbal comprehension with items on vocabulary, similarities, comprehension, information, word reasoning, perceptual reasoning with items on block design, picture concepts, matrix reasoning, picture completion and working memory with requests on digit span, letter-number sequencing and arithmetic (Ryan et al., 2007). The usage of these tests would allow us to undergo the intellectual and developmental categorization in terms as recommended in the Diagnostic and Statistical Manual of Mental Disorders, fourth edition, text revision. This tool is diagnostic standard

for mental health care professionals in the United States and it classifies four different degrees of mental retardation: mild, moderate, severe, and profound. These categories are based on the person's level of functioning. In mild mental retardation, approximately 85% of the mentally retarded population is in the mildly retarded category. Their intelligence quotient score ranges from 50–70, and they can often acquire academic skills up to about the sixth-grade level. They can become fairly self-sufficient and in some cases live independently, with community and social support. In moderate mental retardation, about 10% of the mentally retarded population is considered moderately retarded. Moderately retarded persons have intelligence quotient scores ranging from 35–55. They can carry out work and self-care tasks with moderate supervision. They typically acquire communication skills in childhood and are able to live and function successfully within the community in such supervised environments as group homes. While severe mental retardation, which touched 3–4% of the mentally retarded population, intelligence quotient scores ranged from 20–40. They may master very basic self-care skills and some communication skills. Many severely retarded individuals are able to live in a group home. Finally, the profound mental retardation is found in only 1–2% of the mentally retarded population. Profoundly retarded individuals have intelligence quotient scores under 20–25. They may be able to develop basic self-care and communication skills with appropriate support and training. Their retardation is often caused by an accompanying neurological disorder. Profoundly retarded people need a high level of structure and supervision. But instead of focusing on limitation, the American Association on Mental Retardation has developed another widely accepted diagnostic classification system for mental retardation in which the stress is put on the capabilities of the retarded individual rather than on his or her limitations. The categories describe the level of support required. They are: intermittent support; limited support; extensive support, and pervasive support. To some extent, the American Association on Mental Retardation classification mirrors the Diagnostic and Statistical Manual of Mental Disorders, fourth edition, text revision classification. Intermittent support, for example, is support that is needed only occasionally, perhaps during times of stress or crisis for the retarded person. It is the type of support typically required for most mildly retarded people. At the other end of the spectrum, pervasive support, or life-long, daily support for most adaptive areas, would be required for profoundly retarded persons. The American Association on Mental Retardation classification system refers to the "below-average intellectual function" as an intelligence quotient of 70–75 or below (AAMR, 2000). The patients' evaluation was completed by physical examination to find out any striking or clue thing that could lead to an accurate diagnosis evocation. Information collected about the child's presentation, their medical history and that of their family can be used to focus attention on particular systems or organs during examination. It is important however to carry out a full physical assessment to avoid missing the subtle sign which may be vital. Assessment for dysmorphic features is important. Explaining to parents why appearance is important can be difficult and it is vital to avoid any suggestion of the 'funny looking kid' concept which is thankfully heard very rarely now. It is perhaps easiest to describe the process as the recognition of patterns of appearance that are seen in most children who have the same condition but are not typical of their family. The availability of other family members to compare the affected individual to either in person or as photographs can greatly aid the assessment as can the opportunity to review pictures of the affected child over the years. This is particularly helpful if the young person is approaching adulthood when the classical appearance of many of the dysmorphic syndromes may have altered. The most effective way to record a dysmorphological assessment is by the use of photography.



The introduction of digital imaging has made this much more straightforward. With appropriate parental consent a full face view, right and left lateral views, a full body view and close up views of any specific dysmorphic features are a vital part of the medical record (Wright et al., 2009). The morphological examination must be complete (skull, face, neck, hands, feet, chest, mouth) with an accurate classification and recognition even if it must take into account, of course, individual variations and some number of minor anomalies that can be found in the general population. Clinical examination should be complete: with growth measurement such as weight, height, occipito-frontal circumference, cardiac, abdominal, neurological and bones and joints function, skin, nail and hair presentation, external genitalia aspects. One must call for other specialists helps if needed (dermatology, ophthalmology, ear, nose and throat...). At the ends of this evaluation, the mental retardation could be classified either in syndromic group (association with birth defects, dysmorphism, specific neurological, growth abnormality, sensory deficiency. . .) and non-syndromic intellectual and developmental disability (no other symptoms apart from the developmental delay) (Goldenberg et al., 2010). The next steps in the global evaluation of a child with intellectual and developmental disability is laboratory investigation. Finding a cause for intellectual and developmental disability is important for prognosis, genetic counseling, and, in some cases, for therapy, although it has to be accepted that cause will remain unknown in many cases and that a combination of multiple factors may be more frequent than single causes thus making decision difficult, especially in mild cases. Screening for specific diseases is a highly effective diagnostic method. Whenever a reliable and simple test is available, an effective treatment is possible and the frequency of the target disorders is sufficient. Its major interest lies in the possibility of preventing intellectual and developmental disability. A thorough search for metabolic disorders is important for both therapeutic and genetic reasons and is especially in order when there is a family history of intellectual and developmental disability or neurological diseases, when close parental consanguinity is present, and when the condition is progressive and a history of a free interval of hours to months after birth is elicited. However, many metabolic diseases are now known to be clinically manifest from birth and may be static or very slowly progressive or even be associated with brain and peripheral malformations, so the indications of metabolic investigations have to be extended. The routine use of amino acids and organic acids chromatographies of the urine for diagnosis of metabolic causes of intellectual and developmental disability is not entirely reliable as excretion of abnormal metabolites may be intermittent or require previous loading and rarer metabolites may demand specific techniques. It may be more cost effective to study in detail those cases in which a clinical suspicion is present. A careful clinical examination is essential looking for skin abnormalities, dysmorphism, skeletal anomalies, suggestive of neurocutaneous syndromes, or of genetic dysmorphism syndromes. The latter have become increasingly numerous and their recognition can be considerably aided by available data bases including pictures, such as the London Dysmorphic Data Base or the Possum Data Base. Some of these syndromes (Williams's syndrome or Smith-Lemli-Opitz syndrome) are not detectable by the use of deoxyribonucleic acid or biochemical tests. Neuroimaging studies, especially magnetic resonance imaging, are useful in many cases of severe cases. Magnetic resonance imaging is definitely indicated when epilepsy or neurological signs are present and the frequency of cortical malformations in such cases is well documented. Chromosomal analysis is indicated for cases with clear cut dysmorphism. However, many chromosomal aberrations are not associated with major dysmorphism, and a karyotype is indicated whenever minor peripheral malformations are found. A routine karyotype is often enough, and more specific techniques of deoxyribonucleic acid analysis are essential for confirmation (Aicardi 1998).

#### 4.4 Investigations

Specific tests are often requested when investigating the child or young person with intellectual developmental disability. Following the review of the available evidence in 2006 by McDonald et al., these coming tests could be considered as base line investigation for children with global developmental delay. These include in the first line, karyotype, deoxyribonucleic acid analysis for Fragile X syndrome, urea and electrolytes, full blood count, creatin phospho kinase, thyroid function with thyroid-stimulating hormone, thyroxine and triiodothyronine, urate, ferritin, biotinidase and lead rate. If these tests give no positive result, one could consider second line investigations with metabolic testing taking in consideration family history, consanguinity, regression, coarse facial features, organomegaly. It is completed by blood-lactate, amino acids, ammonia, very long chain fatty acid, carnitine, homocysteine, disialotransferrin, urine-organic acids, orotate, glycosaminoglycans and oligosaccharides. Neuroimaging including magnetic resonance imaging or computerized tomography scan  $\pm$  electroencephalogram could be done if there is any neurological abnormality such as microcephaly or macrocephaly, seizures, focal neurological signs (McDonald et al., 2006). Only few of these could be afforded in our setting. We usually proposed karyotyping since this technique is available in our country. This situation could be considered as an abnormal one comparing to what is done in western. But one must keep in mind that in the whole west Africa sub region and probably in the large sub-Sahara region, there is no where apart from Cotonou, in Benin one could find this cytogenetic technique. One could find some in South Africa, Tunisia, Egypt and Morocco as reported by world health organization in an official document (WHO, 2005). This laboratory unit plays then a regional role by receiving sample from neighboring countries such as Togo, Cote d'Ivoire, Niger, and Burkina Faso. Discussions are going on to include more especially Senegal and Nigeria which are key countries in West Africa in terms of advance in medical practices. In this setting we do a lot in cytogenetics but our skill is limited in molecular genetics which become nowadays important tools in the single gene disorder diagnosis. We only could extract deoxyribonucleic acid and coop with a couple of molecular testing such as beta S mutation in sickle cell anemia prenatal diagnosis and deletion in Azoospermia factor in sterile men with azoospermia (Gangbo et al., 2009). Otherwise, we send our deoxyribonucleic acid extracted overseas through network cooperation. This is a great opportunity that we have established while studying in some European countries especially in France and Belgium. This has assisted us a lot in moving forwards and been able to answer to our colleagues and to the family who claim to be informed on their children condition.

#### 4.5 Etiologies of intellectual and developmental disability

This current survey on intellectual and developmental disability mainly in children put forwards some etiologies. These could be categorized in three groups with inheritable disorders, environmental diseases and unknown condition (Aicardi, 1998). The inheritable aetiologies accounted for almost 25% with Down syndrome taking one out of five cases. The environmental factors were birth asphyxia and jaundice and both of which gave 40% responsibility to neonatal conditions. The remaining cases have no cause and this is best known among people that work on intellectual and developmental disability or simply on mental retardation. But although in 60% of cases of intellectual disabilities the causes are unknown, four categories of factors that can occur before, during, or after birth have been

identified as etiological factors and these include genetic disorders, chromosomal disorders, biological and organic causes, and environmental causes. Actions can be undertaken to alleviate the effect of some of these factors as we will see infra (WHO, 2007). The leading cause we found was indeed birth asphyxia and it is well established that it causes a lot of intellectual and developmental disabilities in children mainly in developing countries (Haider and Bhutta, 2006; Lanwn et al., 2008). This is a world burden and one of the major causes of morbidity and mortality in neonate all over the world (Black, 2010; Lawn et al., 2005; Lawn et al., 2008). A neonate is labeled to be asphyxiated if the following conditions are satisfied: umbilical cord arterial pH was less than 7, Apgar score  $\leq 3$  for longer than 5 minutes, newborn showed neurologic manifestations such as seizures, coma or hypotonia and multisystem organ dysfunction (cardiovascular, gastrointestinal, hematologic, pulmonary, or renal system) could be found. Thus hypoxia or asphyxia should be labeled as a cause of disability and handicap only when the neonate demonstrates the four perinatal findings listed above and in whom other possible causes of neurologic damage have been excluded. In the absence of such evidence, subsequent neurologic deficiencies cannot be ascribed to perinatal asphyxia or hypoxia (Haider and Bhutta, 2006). Some neonates that suffered from birth asphyxia could present symptoms of cerebral palsy but one must keep in mind that intellectual and developmental disability is totally different to the first one even if they can negatively influence mutually (McCullough et al., 2011). Jaundice is also a major environmental cause in this survey and this health condition is frequent in neonate (Lawn 2008; Slusher et al., 2011). It could arise from immunological incompatibility, severe neonatal infection, delay in liver function in premature babies and some enzymatic deficiencies (Kaplan, 2010). This survey showed as that apart from environmental factor that must be tackled if one hopes the reduction of intellectual and developmental disabilities in children; there is a huge contribution of genetic diseases. The very first group of inherited diseases was Down syndrome. This condition was frequent in our setting and forced us to open a special program for the affected children (Alao et al., 2010). Down syndrome is a common cause of developmental disability. There is widespread awareness of the associated physical features and variable learning disability, but possibly less understanding of the wide range of health problems which may also affect those with the syndrome. Increasingly diagnosis of Down syndrome is made antenatally and many affected pregnancies are terminated. Despite the fact that a lot of such pregnancies were terminated there has been no major change in birth prevalence. This is probably because women are now starting their families later, and as incidence of Down syndrome rises with maternal age there are likely to be more conceptions of babies with Down syndrome. Babies born to those women who decide to continue the pregnancy after diagnosis of Down syndrome, together with those diagnosed after birth (false negative, or screening not performed), currently give a live birth rate of 1.08/1000. It is therefore likely that there will continue to be more than 700 babies with Down syndrome born each year worldwide. Paediatricians have a key role in health provision for these children. Whilst some thrive from an early age and are in good health throughout childhood there is, among the group as a whole, an increased risk of congenital abnormalities and a wide range of medical problems. The impact of these problems on general health, growth and development may be even greater than would be expected for other children because of the associated developmental delay and learning disability. Historically, some treatable conditions were thought to be 'part of the syndrome' and left

untreated. There may have been lack of recognition of potential benefits to overall functioning of the child or even discrimination. Today we hope that the health of children with Down syndrome will be monitored as carefully as that of any child, and treatment offered when necessary so that their progress is not hampered by additional secondary but preventable handicap, and that health problems do not prevent them reaching their potential. The cognitive development of children with Down syndrome is characterized by interindividual variability in their cognitive functioning (Tsao and Kindelberger, 2009). The other genetic diseases were represented by some very rare condition even in the developed countries. The first of them was Patau syndrome due to trisomy 13. This condition was seen in an infant that was referred at age of 6 months for psychomotor delay. The parents have experienced a spontaneous first trimester abortion. The actual pregnancy was carried out with no abnormalities and the ultrasonographic exams were normal. Delivery took place at 37 weeks of gestation and Apgar score disclosed 6, 8 and 9 respectively at 1, 5 and 10 minutes. Neonatal resuscitation was conducted for 3 minutes with skin drying, nose, throat suction and oxygen administration. Birth measures showed 3.100 kg for the weight (55<sup>th</sup> percentile), 50 cm for the height (75<sup>th</sup> percentile) and 33 cm for the fronto-occipital circumference (50<sup>th</sup> percentile). Neonatal period was marked by jaundice. Later, the infant suffered from medium otitis, upper airway obstruction and frequent respiratory tract infections. Physical examination at age of 6 months disclosed postnatal growth retardation, psychomotor delay with the lack of sited position, craniofacial dysmorphism with microcephaly, hirsutism, coarse face, flared nostrils, large auricles, macrostomia with enlarged tongue, long and large philtrum, umbilical hernia and brachydactyly. She also showed noisy breathing and kyphosis. Mucopolysaccharidosis type 1 was evoked regarding the facial dysmorphism. The child was referred to cytogenetics laboratory for testing including karyotyping and deoxyribonucleic acid extracted extraction for further analysis. Surprisingly, the karyotype showed full trisomy 13. Parents' karyotypes were normal. She is still alive after 2 years with very poor communication ability. This case reveals the possibility of mixture in causes with birth asphyxia, neonatal jaundice and confirmed chromosomal abnormality (Aicardi, 2000). We also diagnosed one case of Di George syndrome in a boy with intellectual disability. This syndrome is characterised by the association of several malformations: hypoplastic thymus and parathyroid glands, congenital conotruncal cardiopathy, and a subtle but characteristic facial dysmorphology. Velocardiofacial syndrome is marked by the association of congenital conotruncal heart defects, cleft palate or velar insufficiency, facial dysmorphology and learning difficulties. It is now accepted that these two syndromes represent two forms of clinical expression of the same entity manifesting at different stages of life. The characteristics defining these syndromes overlap with those of microdeletion 22q11. The acronym CATCH 22 was proposed to describe the clinical features of microdeletion 22q11 (Cardiac-Abnormal face-Thymus-Cleft palate-Hypocalcemia). The clinical course of the syndrome is mainly determined by the nature of the congenital malformations involved. The hypocalcemia frequently observed in the neonatal period generally disappears, but some children may have persistent hypoparathyroidism, which requires treatment. The velopharyngeal insufficiency often results in nasal speech, even in the absence of cleft palate, and may be associated with language difficulties. Microdeletion in 22q11 is present in 95% of patients. The incidence of the microdeletion in 22q11 in the general population is estimated at 1 in

5000 births. In 10 to 20% of cases, the 22q11 microdeletion is transmitted in an autosomal dominant manner, with one of the parents being a carrier of the microdeletion. However, in the majority of cases, the chromosome anomaly arises *de novo* (Jones, 1997). We did not have yet a molecular confirmation in the case of Aarskog syndrome but the features that were found were relevant to evoke the hypothesis. The boy showed indeed craniofacial dysmorphism with bombing front head, hypertelorism, upturned nose, webbed cheeks, unmarked philtrum and curved linear groove below lower lip. Hands and feet were short and broad with interdigital webbing, clinodactyly and edema of limbs. The stature was short with small scrotum (Taub and Stanton, 2008). In this disease, there is no genotype phenotype correlation regarding the intellectual disability. The affected children with mental impairment are only mildly affected, with learning and behavioural disabilities often confined to early childhood. The majority of these children have a good evolution into adulthood and 'the changing phenotype with age' includes an age-related improvement of mental status. However, the risk for a variety of behavioural disturbances and mild learning difficulties appears to be increased in these children and specific attention to cognitive and behavioural function is needed in young Aarskog syndrome patients. Only a minority of the clinically diagnosed patients carries a mutation in the facio-genital dysplasia 1 gene. The diagnosis of the X-linked Aarskog syndrome needs to be made with care as the spectrum of clinical signs overlaps with that of many different disease entities and, although the phenotype may be impressive, many alternative diagnoses have to be considered (Orrico et al., 2004). We got two cases of mucopolysaccharidosis type I that is also called Hurler syndrome with a severe intellectual disability. Developmental impairment is well known in this storage disease. Neuropsychological manifestations in patients with mucopolysaccharidosis type I may arise from primary glycosaminoglycans accumulation in the central nervous system or be caused by deposits in adjacent structures such as the meninges and bone structures. In general, patients with this condition develop normally or have only mild developmental delay in the first year of life. In severe Hurler syndrome, developmental delay is observed between 12 and 24 months of life, chiefly in the speech realm, with subsequent progressive cognitive and sensorial deterioration, most markedly in visual and auditory areas. The mental, motor, and behavioral status of patients can be monitored with developmental scales and intelligence quotient tests. The combination of these assessments can furnish relevant information on intellectual deterioration and clinical evolution in patients. Choice of assessment instruments (psychometric tests) should be based on chronological age and the patient's visual, auditory, and motor abilities (Martins et al., 2009). The last genetic diseases we succeed in catching were two cases of Apert syndrome. The typical facial features of this syndrome include a characteristic break in the eyebrows, ocular hypertelorism, downslanting palpebral fissures, and thin upper lip with a trapezoid or tented appearance. Head shape can be extremely turribrachycephalic with moderate to severe midface hypoplasia. Initially, there is a wide calvarial defect from the posterior fontanel to the glabella, and the anterior portion of the defect is sometimes described as an "encephalocele," which is a misnomer because bony obliteration eventually occurs. The malformations of the central nervous system seen in this disorder are numerous, including hydrocephalus, ventriculomegaly, megalencephaly, gyral malformations, and defects in the corpus callosum, septum pellucidum, hippocampus, and cerebral cortex. Cleft palate and hearing loss because of fused ossicles are also observed. There are varying

degrees of developmental delay. Generally, intelligence quotient correlates inversely with intra cranial pressure however, the developmental delay may be unrelated to the increased intra cranial pressure because of the fact that the large midline skull defect and widely patent fontanels do not give rise to intra cranial pressure early in development. Skeletal problems are severe and multiple, including bony syndactyly of the hands and feet with sparing of the thumb, giving the impression of a "mitten hand." Fused cervical vertebrae (68%, usually C5-C6) and elbow ankylosis are seen. Other congenital anomalies can occur such as cardiac (10%) and genitourinary (9.6%) defects. Two hotspot mutations in Fibroblastic Growth Factor- Receptor 2 gene (S252W and P253R), account for the majority of cases (71% and 26%, respectively). Some genotype-phenotype associations have been suggested (for example the severity of the syndactyly with the P253R and the presence of cleft palate in S252W). A paternal age effect in de novo mutations in Fibroblastic Growth Factor- Receptor 2 gene has been conclusively shown at the molecular level in Apert syndrome. It has been hypothesized that mutations in Fibroblastic Growth Factor- Receptor 2 gene may convey an advantage in sperm because the Fibroblastic Growth Factor/Fibroblastic Growth Factor- Receptor pathway is known to be important in maintaining and initiating spermatogenesis (Kimonis et al., 2007). There is normally no neurodevelopmental disability in Marfan syndrome. Apart from behavioral disturbances and some difficulties to rule out right diagnosis, no mental problem is related to this condition. Indeed, the large variation in phenotypical expression of clinical features can make diagnosis difficult. Large variation can even be encountered between affected individuals of the same family. Overlap of clinical features with other Marfan-like disorders also requires care when diagnosing a new case of Marfan syndrome. The differential diagnosis includes homocystinuria, Beal's syndrome (congenital contractural arachnodactyly), Ehlers-Danlos syndrome, Stickler syndrome (hereditary arthro-ophthalmopathy), Klinefelter syndrome, familial mitral valve prolapse syndrome, multiple endocrine adenomatosis and X-linked mental retardation with marfanoid habitus (McBride and Gargan, 2006). So we cannot precisely distinguish the real cause of intellectual disability associated to our Marfan syndrome case even if it was very mild one. This could arise from cardiac dysfunction with neurological impairment. Most of our aetiologies were reported in previous survey either in national based data collection or hospital survey (Masri et al., 2010; Lin et al., 2009; Wu et al., 2010). But one could notice the absence of data on Down syndrome which is probably ruled out during pregnancy screening. Regarding our aetiologies, we could do more if we have access to some screening techniques such as metabolic testing, fluorescence in situ hybridation, comparative genomic hybridation, brain computed tomography scan or magnetic resonance imaging. Then we could report more causes such as the one that were found in a recent Jordanian studies. Indeed, they succeeded in collecting in a population of 229 children with global developmental delay, a large range of causes from cerebral palsy (31.4%), metabolic disorders (6.5%), other single gene disorders (5.2%), brain malformations (3.0%), chromosomal disorders (2.6%), autism (5.2%) and undetermined (55.5%) (Masri et al., 2010).

#### 4.6 Management

Management of the child with mental retardation or intellectual and developmental disability, as with any chronic condition, should not only focus on the child and his

condition, but also on the family. The family is the child's best resource. Supporting the family and ensuring its emotional and physical health is an extremely important aspect of overall management. The four major aspects of caring for children with intellectual and developmental disability include health (growth, developmental, and behavioral surveillance, and mental and dental health); developmental and educational interventions; community integration through social and recreational activities; and special considerations in adolescence and transition to adulthood. Management of children with mental retardation or other intellectual and developmental disability varies depending on presence or absence of a known syndrome and on the severity of the disability. The advantages of the etiological diagnosis is the availability of a standard management "protocol" as has been developed for some conditions like Down syndrome. These protocols are useful in guiding the clinician in surveillance and screening strategies for comorbid and secondary conditions. Children with mild mental retardation or other intellectual and developmental disability are more likely to have idiopathic condition and to be healthy. Thus, health care may vary little from typically developing children. Their degree of "diversity" may be somewhat more obvious in educational and community settings. For these children, transition to adulthood may require extra effort but they are usually able to live independently and may marry, have a family, and work in a competitive job. On the other hand, children with severe intellectual and developmental disability are more likely to have a known etiology complicated by characteristic medical, behavioral, and psychiatric comorbidities that challenge health management and may shorten life span. These individuals will usually require more intense special education as well as additional supports to facilitate community integration and transition to adulthood. As adults, they are less likely to live independently, marry, and parent children. Regarding health interventions, the first step is breaking the news in a sensitive, compassionate, and culturally appropriate manner. When breaking the news, it is important to emphasize the child's strengths as well as the deficits. It is also important to be realistic without taking away hope. As with any child, children with intellectual and developmental disability will benefit from comprehensive health care. In children with delays, it should also be developmentally appropriate. Parent-professional partnerships, built on a foundation of mutual responsibility and trust, are also important. The quality of these partnerships have been rated as one of the most important aspects of medical care by parents of children with chronic conditions such as intellectual and developmental disability. Comprehensive health care for any child should address growth, developmental and behavioral surveillance, anticipatory guidance and safety counseling, as well as traditional medical and dental care. In some children, psychiatric and therapeutic (physical, occupational, and speech therapies) services may be needed. Delivery of medical and dental care to a child with mild mental retardation may be very similar to that of children of normal intelligence. Anticipatory and safety counseling should be modified to reflect the child's mental age rather than his chronological age. Providing care may be somewhat more challenging in children with severe levels of intellectual and developmental disability as they are more likely to be nonverbal and to have comorbid medical, behavioral, and psychiatric conditions. Physicians may be required to spend extra time and effort in communicating and coordinating care with subspecialists, school personnel, and community agency staff. Attention must be paid to these following comorbidities in children with intellectual and developmental disability, especially in those with more severe degrees: behavior disorders, psychiatric disorders, seizures, sensory impairments with hearing and

vision impairments are also more common in children with intellectual and developmental disability, motor impairments, sleep disorders, gastrointestinal symptoms, autism or autistic-like behaviors. Developmental and educational interventions are given according to the child's age. In either case, services should begin as soon as the delay or deficit is recognized. Families will need additional patience and persistence when raising a child with intellectual and developmental disability. Early intervention program should be offered if possible. If the child is not toilet trained eligibility for school services and admission may be denied. Children showing intellectual and developmental disability can attend pre, primary and high school with special supportive condition speech, occupational, and/or (but less frequently) physical therapy. When service delivery is not feasible in the regular classroom due to severe degrees of cognitive impairment or behavior problems, the child might attend a "resource classroom" for one or more academic subjects. Teenagers or adults can be offered competitive employment with unskilled, semiskilled, or in some cases, even skilled duties, supported employment with specific duties through coaching, sheltered employment with works under constant supervision in a segregated setting. Community integration could be achieved through recreational, scouting, and social activities outside of the school arena. These activities help promoting community integration over the life span. As is the case in the educational arena, absence of maladaptive behaviors may be more important to the child's successful inclusion in these activities than level disability. All persons, including individuals with disabilities, benefit from recreation and leisure activities. As with typically developing children, those with intellectual and developmental disability will have particular interests and talents that are unique to the individual. Additionally, some syndromes are associated with unique abilities; for example, girls with Rett syndrome often demonstrate a strong affiliation for music, so much so that new information presented within a musical context is more easily learned than through traditional verbal means (Johnson et al., 2006). Our cases had benefited from symptomatic treatment like what is generally given to children. This was related to respiratory tract, ear, nose and throat and digestive infections and malaria. These are the common diseases in children especially in less than five in our setting (Black et al., 2010). Apart from these, special attention was given to each group to rule out known complications. For example, in the group of Down syndrome, systematic investigations were conducted to exclude many treatable or preventable complications. It is generally recommended that children with Down syndrome must be offered regular medical review by a paediatrician throughout childhood. This may be via a hospital department, child development centre, or community paediatric service. The type of service offered will vary according to individual need as well as local service organization. These children could experience cardiac disorders such as congenital malformations, acquired valvular dysfunction; orthopaedic difficulties such as cervical spine disorders, hip subluxation/dislocation, patellar instability, scoliosis, metatarsus varus and pes planus; ear, nose and throat problems like conductive hearing loss, sensorineural hearing loss, sleep related breathing disorders and chronic catarrh; ophthalmic disturbances such as refractive errors, blepharitis, nasolacrimal obstruction, cataracts, glaucoma, nystagmus, squint and keratoconus; gastrointestinal disorders such as congenital malformations, feeding difficulties, gastro-oesophageal reflux, Hirschprung's disease and coeliac disease; endocrinal problems like hypothyroidism, hyperthyroidism and diabetes. Some could showed more like immunological troubles such as immune dysfunction,



autoimmune diseases; haematological disorders especially leukaemia, polycythaemia, macrocytosis and leucopenia; dermatological finding with dry skin, folliculitis, vitiligo and alopecia. Neuropsychiatric problems apart from a real developmental delay may include infantile spasms and other myoclonic epilepsies, autistic spectrum disorder (Charleton et al., 2010). Our education rate was too low. Our schedule in this sector is to try to help children attending ordinary school with four years in preschool instead of two and two or three years in each form of primary school. The most advanced of them was in primary 2. Education is important since these children could progress if they are well trained according to their specific capacities (Mancini et al., 2000; Kozulin et al., 2010; Giaouria et al., 2010). Supportive care was offered to some of our children. The most used were physiotherapy and speech therapy but all of them could not receive the service due to lack of finance. Indeed, the patients were supposed to cope with the treatment fees since there is no general or universal insurance system in the country. Some news techniques such as chiropractic treatment are available as complement to other supportive care (Cuthbert and Barras, 2009). The six cases of autism spectrum disorders had no specific aetiology since we did not have occasion in searching them (Betancur 2011). But they were seen in psychiatric consultation and their condition was confirmed through outpatient system even if some could normally be kept in hospital for more accompaniments (Tremblay et al., 2010; Friedman et al., 2011). They also received speech therapy and some could show more communication ability because we took as example the phonological awareness of children with Down syndrome (Lemons and Fuchs, 2010). Family could help in the treatment program (Embregts, 2009). Nurses could participate in the management of children with intellectual and developmental disability if they are well trained and aware of this issue (Hicks and Clark 2011). We also need to protect young people with intellectual and developmental disability against abuse, especially in female subgroup (Doughty and Kane, 2010).

#### **4.7 Prevention**

Prevention will be the most worthy activity since there is no real etiologic or curative treatment when the disability is present. A lot of strategies are promoted. These generally speaking included supplementation of diet by iodination of salts or folic acid in bread (in 67.1% of countries); programmes for prevention of alcohol or drug abuse during pregnancy (61.6%); genetic counselling and prenatal testing (61.0%); and tests to detect phenylketonuria, lead, or hypothyroidism (57.5%). These strategies were more common in high-income countries than in low-income countries (WHO, 2007). We have here to focus on the finding causes. Birth asphyxia could be prevented by increasing the births attended and by promoting community participation in this activity (Darmstadt et al., 2009). Since after decades of intervention in health centers, there is no real change in this sector, actions are currently directed towards community participation in Emergency obstetric and neonatal care but one must not ignore the huge role of behavior changing that is needed. Indeed, behavior change is a critical and fundamental determinant of newborn survival. It is not only the primary component of preventive interventions but a necessary complement of more downstream therapeutic interventions, thus providing the "last mile connectivity" between the existing evidence base and impact on newborn survival at a community level. It is time that we begin to address the gap between "what works" and "how to make it work." Behavior change can contribute substantially towards minimizing

this gap, and, therefore, it is critical to systematically understand the determinants of newborn care behaviors, their underlying sociocultural context, and leverage this knowledge to develop mechanisms for effectively changing behaviors at a population level (Kumar et al., 2010). But overall, newborn resuscitation needs to be carried out in all the settings where asphyxiated babies are born, including: community or domiciliary settings for home births; rural health centers/midwifery stations, where attendants with basic resuscitation skills might be available; district-level facilities where staff are available but skills vary; and urban referral and tertiary care centers. Individuals at all levels require training and seldomly used skills need to be maintained so that, when required, resuscitation can be carried out efficiently and effectively. Simple resuscitation techniques include: positioning, drying, and keeping the baby warm; assessing the heart rate, color, and respirations; recognizing the need for, and administering, assisted ventilation with a bag and mask or tube and mask. These maneuvers can be carried out with simple equipment and appropriate training (Singhal and Bhutta, 2008). Neonatal jaundice deserves more attention. It could be prevented by a regular pregnancy follow-up, delivery in a good hygienic condition and a systematic screening to rule it out during maternity staying. Early discharge must be abandoned. Application of ten basic rules will help preventing hyperbilirubinemia and poor neurodevelopment outcome. These include breastfeeding promotion, jaundice protocols identification establishment, total serum bilirubin or transcutaneous bilirubin measurement on infants jaundiced in the first 24 h, awareness that visual estimation of jaundice can lead to errors, particularly in darkly pigmented infants, interpretation of bilirubin levels according to the infant's age in hours, caution with infants <38 weeks, particularly if breastfed, who have a high risk, risk assessment performing on all infants prior to discharge, parents written and oral information about jaundice, appropriate follow-up based on time of discharge and risk assessment and newborns with jaundice treatment, when indicated, with phototherapy or exchange transfusion (Maisels, 2010). More effort should be made to avoid isoimmunization and Glucose 6 phosphate deshydrogenase deficiency (Geaghan, 2011; Olusanya and Slusher, 2010). Down syndrome could be prevented by the implementation of antenatal diagnosis. The following screening tests for fetal Down's syndrome were evaluated: measurement of first-trimester nuchal translucency alone; first-trimester serum screening alone (pregnancy-associated placental protein-A and free-beta subunit human chorionic gonadotropin were measured); first-trimester combined screening (nuchal translucency plus pregnancy-associated placental protein-A and free-beta subunit human chorionic gonadotropin); second-trimester quadruple screening (alpha-fetoprotein, total human chorionic gonadotropin, unconjugated estriol, and inhibin A); independent sequential screening (the results of combined screening were provided to the patient in the first trimester, and the results of quadruple screening in the second trimester, with both risks calculated independently); stepwise sequential screening (the results of combined screening were provided in the first trimester, and the results of quadruple screening in the second trimester; the risk in the second trimester was calculated with inclusion of the marker levels measured in the first trimester); serum integrated screening (pregnancy-associated placental protein-A was measured in the first trimester, and the results were not provided to the patient; quadruple markers were measured in the second trimester, and the risk in the second trimester was calculated with inclusion of the marker levels measured in the first trimester); and fully integrated screening

(identical to serum integrated screening with the addition of first-trimester measurement of nuchal translucency) (Malon et al., 2005). Community should be associated in the process of better maternal and neonatal health condition. Since it there is evidence that community mobilization is an effective method for promoting participation and empowering communities among a wide range of other non-health benefits (Rosato et al., 2008).

#### 4.8 Social considerations

Children with intellectual and developmental disability receive a little attention from the society because they are not considered as having disability. They are seen some times as divinity with worship activities or as devil and could some time receive sacrifice or purification. This sacrifice could lead to the child death by throwing him to “mamywater” or leaving him in a corner with no food or in the bush.

#### 4.9 Perspectives

Perspectives in child with intellectual and developmental disability should be seen in attempt to improve management. Diagnosis should be improved specially regarding genetic diseases by more access to techniques like fluorescence in situ hybridation our comparative genomic hybridation array through network cooperation. It is well known that comparative genomic hybridation array can upgrade the rate of positive finding in this population (Jaillard et al., 2010). Treatment must be arranged with urgent creation of a dedicated child protection service for children with intellectual and developmental disability (Shannon and Tappan, 2011). More researches are needed to well understand this issue especially at community level.

### 5. Conclusion

Intellectual and developmental disability is not rare in Benin especially in children. General diagnosis tools were available and need to be strengthened. They were provoked most time by birth asphyxia, jaundice and some genetic condition like Down syndrome. They could be managed through medical basic follow up with supportive cares. Efforts are needed to break financial barrier towards ethological investigations, specialized consultations and occupational activities such as school attending. More research are awaiting is waiting to understand the issue at community level and find out parents consideration and needs.

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