

**Collective Expert Report**

# **Deficiencies and handicaps of perinatal origin**

## **Screening and management**

**Summary**

**Inserm**

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This document presents the summary and recommendations of the expert working group brought together by Inserm within the framework of the collective expertise procedure to answer the questions raised by the Office parlementaire d'évaluation des politiques de santé (OPEPS) (parliamentary office for the evaluation of health policies) concerning deficiencies and handicaps of perinatal origin. It is based on scientific data available during the first half of 2004. Approximately 700 articles and documents form the documentary basis of this expert report.

The Inserm collective expertise centre has co-ordinated this collective expert report with the Département animation et partenariat scientifique (Daps) (department of guidance and scientific partnership) for preparation of the dossier and with the documentation section of the Département de l'information scientifique et de la communication (Disc) (Department of scientific information and communication) for the bibliographical research.



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# Foreword

The origin of more than half of child deficiencies and handicaps could well date back to pregnancy or birth. Two fundamental causes are severe or moderate prematurity and foetal suffering. An increasing number of children are born very prematurely in France every year and their survival rate is improving. Thus the number of children at risk of developing deficiencies or handicaps is also on the increase. Considerable progress has been achieved in recent years to prevent the principal risks associated with both these causes.

The guidance law of 30<sup>th</sup> June 1975 in favour of handicapped people, soon to be replaced by a new law, highlights the national obligation with regard to preventing and screening for handicaps, care approaches, education, training and professional guidance. It has been introduced by the special education departmental commissions (SEDCs) that are skilled in assessing the extent of invalidity, the allocation of financial assistance and guidance in the various structures concerning children between 0 and 20 years of age. In applying this law, the decree of 15<sup>th</sup> April 1976 created early medico-social action centres (EMSACs), located at the interface between the sanitary and medico-social sectors as leading centres in all of the devices used for screening purposes and the early management of children under 6 years of age. Beyond this age, the official guidelines insisted on the co-ordination of policies in order to ensure continuity of education as young infancy is considered to be a crucial period for a child's emotional and cognitive development.

Despite the regulatory provisions in place, measures for optimising screening are still inadequate, both for the general population and risk groups and for organising an early, individualised management approach for children at risk or suffering from deficiencies of handicaps of perinatal origin.

The parliamentary office for the evaluation of health care policies (OPEPS) wished to make use of the Inserm collective expertise procedure to evaluate the knowledge of devices, programmes and policies implemented at international level and in France with relation to the management of children at risk or suffering from deficiencies or handicaps of perinatal origin.

To respond to this request, Inserm set up a group of 14 experts specialising in the fields of obstetrics, neonatology, infantile resuscitation, neuropaediatrics, epidemiology, health economy, physical medicine and rehabilitation, child psychiatry and education.

The experts worked on the basis of the following set of questions:

- What is the prevalence of deficiencies and handicaps? How has this changed over the last few decades? What are the reference classifications concerning handicaps? What is the definition of a handicap of perinatal origin?
- What is the relative proportion of handicaps of perinatal origin taking handicaps as a whole? What are the different types of deficiencies or handicaps of this origin?
- What are the main perinatal events involved in deficiencies or handicaps? What are the upstream risk factors that can be prevented?
- What tools are available for the early screening of cerebral lesions?
- What is the time frame for screening during the first three years of life, depending on the type of deficiencies?

- What treatments and early interventions have displayed a certain efficacy in preventing deficiencies and handicaps?
- What are the management methods employed for each type of deficiency?
- What is the interest of economic studies relating to screening and management within the scope of decision-making?
- What screening and management devices are available in France and other countries? How are families integrated in the various approaches? How are institutional responsibilities divided in France?
- What is the French experience in terms of approach/device networks?
- During 5 working sessions, the expert group analysed over 600 scientific and medical publications and numerous reports and drafted a summary of these studies together with recommendations for action in public health and research recommendations.

## Summary

Considerable progress has been achieved in the field of obstetrics and neonatology in the last thirty years. This was initiated during the 1970s by the national perinatal programme (1970-72), the aims of which were to reduce the number of pregnancy- and labour-related deaths and handicaps. The principal measures introduced enabled improvements to be made in terms of the equipment available in establishments attending pregnant women and the neonatal resuscitation departments, staff training, prenatal monitoring and labour. Later, within the scope of the perinatal plan (1993-2000), the establishments treating pregnant women and infants at birth completed the range of existing equipment. At the same time, advances were made in the management of high-risk populations: establishment of networking, the referral of high-risk women to maternity suites having a neonatal resuscitation department, the circulation of new treatments (antenatal cortico-therapy and surfactants), and the development of neonatal resuscitation. All of these measures were accompanied by a substantial decrease in perinatal mortality rates, which fell from 21 per 1,000 births in 1972 to 7 per 1,000 births in 1998, and the neonatal mortality rate, which, in turn, fell from 14 per 1,000 live births in 1969 to 3 per 1,000 in 1997. A decline of 25-35% in the neonatal mortality rate in premature infants and a decrease of 30-55% in big, premature babies have been recorded over the last fifteen years.

These trends have made it necessary to take new indications into account in order to evaluate perinatal management, especially regarding conditions of survival in children and the onset of a handicap (cerebral, motor infirmity, auditory and visual deficiencies, intellectual deficiencies and psychiatric disorders, etc.). Although investigations into infant handicaps have gradually been imposed, numerous questions are still being asked. These questions concern the definition of the handicap and its specific features in infants. They also focus on the sources of information available in France. In fact, even though the 1970 national perinatal programme explicitly refers to handicaps, there was no means of measuring these at the time. Since then, studies have been carried out in handicapped infants but only to a minor extent in France. We will, therefore, see to what degree they cover an assessment of principal deficiencies, how these advance and their aetiology.

The orientation law of 30<sup>th</sup> June 1975 in the handicapped persons' favour incorporates a series of devices. The new law, which will come into force on 1<sup>st</sup> January 2005 following parliamentary inspection, includes the creation of a compensation law to facilitate the management of human and technical expenditure corresponding to the needs of every handicapped person by various hospitals and institutions. The draft law also aims at improving the educational integration of handicapped children. It outlines the principle of educating such children in the establishment located closest to the parental home with specialist establishments and departments intervening to complement this approach. Action plans should lead to the creation, between 2007 and now, of places in establishments and departments for both children and adults and to provide specific solutions to the requirements of persons suffering from severe handicaps (autistic patients and patients presenting with several handicaps, etc.).

Analysis of national and international data regarding the prevalence, aetiological or risk factors, screening methods and early intervention together with organisation and management programmes, must contribute to the essential knowledge needed in order to implement public policies.

## The International Classification of Diseases (ICD) and the International Classification of Function, handicap and health (ICF) are the two reference classifications.

The International Classification of Diseases (ICD), which was created in 1948 under the direction of the WHO, is the principal official, terminological reference in medicine and epidemiology on an international level. It classifies health problems into diseases, disorders, lesions and traumas. This classification, however, based on a biomedical model, does not allow assessment of the chronic consequences of a trauma or a disease. The International Classification of deficiencies, Incapacities and Handicaps (ICH1) was established in 1980 with an experimental classification statute. This classification considers a handicap as an individual phenomenon and describes the cause-effect relationship in a linear fashion: the disease triggers an organic and functional deficiency – a lesion-related aspect –, which leads to an incapacity in terms of a person's behaviour and activities – a functional aspect – that, in turn, leads to a disadvantage (handicap) – the situational aspect.

The context of the subject's life is, however, not apparent to the ICH1. Between 1996 and 2000, six temporary classifications were co-ordinated by the WHO and the social handicap model was extensively imposed.

In 2001, the WHO General Assembly adopted the International Classification of Function, handicap and health (ICF) as a new tool for defining the state of health of the population and, in particular the prevalence and significance of debilitating situations.

The International Classification of Function refers to three aspects of the handicap: the deficiency, the limitations in terms of activities and participation restriction. Deficiency affects an organ with regard to its function or structure. The limitation in terms of activities and the participation restrictions refer to the difficulties that a person may encounter when carrying out an activity or adopting a social role. The ICF is thus organised into two sections, each comprising two components: the first deals with function and the handicap in question (organic functions and anatomical structures, activities and participation) whilst the second concerns contextual factors (environmental and personal factors). Each component can be expressed either positively or negatively. These components are sub-divided into fields within which are categories (classification units) at two, three or four levels.

### General survey of the International Classification of Function (ICF, WHO, 2001)

	Part 1 Function and handicap		Part 2 Contextual factors	
Components	Organic functions and anatomical structures	Activities and participation	Environmental factors	Personal factors
Fields	Organic functions Anatomical structures	Fields of life (tasks, actions)	External factors affecting function and handicap	Internal factors affecting function & handicap
Diagrams	Changes in organic functions (physiology) Changes in anatomical structure	<i>Capacity</i> to carry out tasks in a standard environment <i>Performance</i> to carry out tasks in a real environment	Impact (facilitator or obstacle) of the physical or social reality or attitudes	Impact of personal attributes
Positive aspect	Functional and structural integrity	Activity Participation	Facilitators	N/A
Negative aspect	Deficiency	Limitation of activity Restricted participation	Barriers Obstacles	N/A

This is a framework classification that allocates a qualitative code defining the importance of the level of health or the severity of the attack (e.g. D166 expresses the reading performance). In the “activities and participation” component, the diagrams distinguish between two qualitative codes: the capacity code, which characterises a person’s aptitude to carry out a task or action in a standard environment, and the performance code, which describes that the person in question is capable of dealing with the context of everyday life. The environmental framework can thus help to limit a person’s activity and his/her participation. The discrepancy between capacity and performance reflects the difference in impact between a standard environment and a uniform environment. The ICF proposes a multi-dimensional vision in which environmental factors play a key role.

This classification can be used at different levels. Everyone, depending on his/her profession, can fit such or such a component. The “organic functions and anatomical structures” component is more specifically informed by the health care professionals whereas the “activities and participation”, “environmental factors” and “personal factors” components are informed by educational and social professionals.

In children, the deficiency component is the most used because it is known by care teams, and it is well described in the literature and can be measured more easily than limitations in terms of activities or participation restriction for which the measuring tools are rare or even inexistent in children.

The literature refers to other approaches or handicap classifications relating to infant/child handicap. Generally developed in the United States, these classifications are essentially pragmatic. They are aimed at identifying the requirements of handicapped children and their families in order to assess the cost and generate assistance. They are not necessarily appropriate for identifying the perinatal origin of the handicap or aetiological approaches. Some authors emphasise the interest of non-category methods that analyse the consequences of various disorders of a medical, behavioural or intellectual performance nature considering that these methods are better suited to provide indications to the departments and organisations in charge of handicapped children.

## **The cause of approximately 50% of deficiencies and handicaps could occur during the perinatal period**

The definition of a handicap of perinatal origin is not unique. For some authors, the handicap of perinatal origin may be defined as a handicap, the causal event of which occurs during life in the uterus after 22 weeks of amenorrhoea (according to the WHO) and up to 8 days or 28 days after birth, depending on whether or not the authors count the peri- and neonatal periods. More often than not, it is only following the elimination of other causes that handicaps are listed, the perinatal origin of which is feasible.

Approximately 20-35% of severe infant/childhood deficiencies are of antenatal origin (abnormal morphogenesis, chromosome aberrations and genetic abnormalities) and between 5 and 10% of severe deficiencies are post-neonatal associated with trauma and, more rarely, an infection or a tumour. Thus, using a process of elimination, it is estimated that 55-75% of deficiencies could originate during the perinatal period. A cautious approach must, however, be adopted when interpreting this aspect because a perinatal risk factor is involved (prematurity, hypotrophy and asphyxia at birth) or a cerebral abnormality of peri- or neonatal origin (anoxo-ischaemic encephalopathy, periventricular leucomalacia and intraventricular haemorrhage) occurs in only 15 to 45% of cases.

These facts highlight the extent of the difficulties involved in trying to establish the origin of handicaps. This particularly affects children in whom no causal or risk factor is likely to account for the onset of the handicap as well as those presenting with a risk factor (prematurity, hypertrophy and asphyxia at birth) without it being formally possible to establish a cause-effect relationship.

### **The part played by perinatal causes varies depending on the type of deficiencies: motor, sensory, intellectual and psychiatric**

According to studies, motor deficiencies include motor infirmity of cerebral origin (MICO), which refers to a motor disorder, the origin of which can be traced back to the perinatal period or cerebral palsy, which signifies permanent motor disorders linked with a non-advancing, cerebral lesion, without pre-judging its origin. These also include deformities affecting the central nervous or musculo-skeletal systems as well as degenerative or hereditary diseases.

Intellectual deficiencies are defined by an intellectual quotient (IQ) of less than 70. According to the WHO agreement, children with an IQ of between 70 and 85 are in a limit zone whilst those with an IQ of 50 to 70 present with moderate mental retardation and children with an IQ of less than 50 are classed as severely or deeply mentally retarded. Intellectual deficiencies also include trisomy 21 regardless of the severity of the mental retardation (but that is not considered as being of perinatal origin).

Auditory deficiencies refer to a bilateral loss exceeding 70 decibels (dB) before correction and visual deficiencies to a visual acuity of less than 3/10 in the best eye (after correction).

Mental disorders occurring between 0 and 3 years of age are mainly autism and other invasive disorders affecting development.

Finally, numerous handicaps associate a motor deficiency with a severe or profound mental deficiency resulting in extremely restricted autonomy and the possibilities of perception, expression and relations.

The role of perinatal causes varies according to the type of deficiencies. For instance, it seems higher in the case of motor deficiencies than for intellectual and sensory deficiencies.

#### **Distribution of deficiencies in France according to their origin (as %), according to RHEOP 1980-1991 (n = 1,360 cases)**

	Proven prenatal causes	Proven post-natal causes	Probable perinatal/ neonatal causes: presence of risk factors		Undetermined causes
			3 simultaneous factors	1 or 2 factors	
Motor deficiencies	32	15	6	19	28
Intellectual deficiencies	52	5	1	11	31
Sensory deficiencies	35	6	2	11	46
Psychiatric disorders	7	3	0	10	80

RHEOP: Register of infant/ child handicaps and perinatal research institute

In France, according to the data listed in the Register of infant/child handicaps and the perinatal research institute (RHEOP), the role of risk factors of perinatal origin in motor deficiencies is twice that for other deficiencies.

**The prevalence of severe child deficiencies in the general population is approximately 1%**

Regardless of the definitions considered and the methodologies employed, most of the epidemiological surveys conclude that the global prevalence of severe deficiencies is approximately 1% in children. It may reach 2% if moderately severe deficiencies are taken into account. However, data relating to the prevalence of handicap depends on the age of the child in question. On average, it takes one or two years to identify a motor or neuro-sensorial deficiency or incapacity, three or four years to detect a fine motor disorder and often longer than that in order to identify a disorder involving cognitive functions. The prevalence will therefore be higher in an older population. Prevalence data are generally set for children in the 5- to 7-year age bracket.

The prevalence of motor handicap (cerebral palsy) is estimated in the population registers of western countries to be between 1.5 and 3 per 1,000 infants born alive.

**Prevalence of principal deficiencies in the general population, taking all causes into account, and based on cohort registers and studies conducted at an international level**

	Prevalence per 1,000 live births
Motor deficiencies (cerebral palsy)	1.5 to 3 (excluding post-natal deficiencies)
Visual deficiencies	
Visual acuity of less than 3/10	0.20 to 1.81
Blindness	0.10 to 0.5
Auditory deficiencies	
Loss fixed at 40 dB	1.1 to 1.3
Bilateral loss exceeding 70 dB	0.4 to 0.7
Intellectual deficiencies	
IQ ranging from 50 to 70	5 to 80
IQ < 50	3.8
Psychiatric disorders (autism and other IDD)	2 to 6

IDD: invasive developmental disorders

According to a recent review of the international literature, almost 3% of children will be moderately mentally retarded (IQ ranging from 50 to 70) but substantial variations can be seen between the studies (0.5% to 8%). As regards severe mental retardation (IQ < 50), the average incidence is estimated to be 3.8 per 1,000 children based on the overall findings of the studies. The heterogeneity between studies is less significant than for moderate mental retardation.

As regards sensorial deficiencies, the prevalence of visual deficiencies ranges from 0.20 and 1.81 per 1,000 live births in Europe and the United States. In the case of blindness, the prevalence varies from 0.10 to 0.50 per 1,000 live births in Europe and the United States. Finally, the prevalence of auditory deficiencies ranges from 1.1 and 1.3 per 1,000 live births for a threshold set at 40 dB between 0.4 and 0.7 per 1,000 in the case of the most serious forms (auditory loss > 70 dB).

Based on numerous international studies (including 3 French investigations), the average prevalence can be estimated at 1 per 1,000 children from 0 to 19 years of age. Taking all of the

forms of invasive developmental disorders into consideration, the average prevalence is close to 3 per 1,000.

The fluctuations in terms of handicap prevalence observed in the literature reflect a better recording of the handicap, changes in medical practices and handicap definition. Furthermore, comparisons of care strategies adopted by teams in the different countries show that the number of children to be looked after, their survival and neurological fate can, to a large extent, be modified by ethical, cultural, social and organisational choices (access to care).

The changing trends observed in recent years in several countries are, nevertheless, convergent. Taking all births into account, the prevalence of deficiencies has stagnated or even increased between 1970–1975 and from 1985–1990.

### **Half of the severe childhood deficiencies or handicaps affect children born full-term or close to term**

The presentation of prevalence rates of handicaps of perinatal origin and changes in these rates can generally be clearly distinguished between infants born full-term and premature infants. Studies focusing on the fate of infants born full-term mainly come from the population registers whereas the studies concentrating on the fate of premature infants or infants with a low birth weight arise especially from cohort studies conducted in reference hospital centres (United States, Canada and Japan) or are based on regional surveys (Europe and Australia).

In premature infants, the deficiency rate is far higher than in that observed in infants born full-term, namely of the order of 3 to 10% for the cohort of very premature babies following less than 33 weeks of amenorrhoea (AW) or even higher in the lower gestational age brackets.

#### **Prevalence of cerebral palsy in children born full-term and in very premature babies (according to Nelson, 2002)**

	Very premature infants (or < 1,500g)	Newborn infants born full-term
% of births	1 to 2%	90 - 95%
Frequency of cerebral palsy per 1,000 live births	55 - 100	1
Role in prevalence of cerebral palsy	Approximately 25%	Approximately 50%

The probability of seeing a deficiency or handicap develop is, overall, inversely proportional to the gestation period or birth weight. The prevalence of handicaps must, therefore, be considered per week of gestational age.

#### **Prevalence of cerebral palsy according to gestational age**

Gestational age	Prevalence (per 1,000 live births)
Before 28 AW*	80 - 95
28 - 31 AW	50 - 60
32 - 36 AW	3 - 17
At term	1

\* Amenorrhoea weeks

A marked increase in the prevalence of cerebral palsies was observed between 1975 and 1985 in very premature infants, especially for the more immature amongst them (born before 28 AW). Since then, prevalence has stagnated or even decreased in these infants. In infants born between 31 and 36 AW, the prevalence of cerebral palsy fell or stagnated between 1980-85 and 1990-95. Finally, no changes were observed in infants born full-term during the same period.

**Comparison of the prevalence rates of the principal deficiencies in the general population, in very premature infants and infants born full-term according to the international data available**

	Prevalence in the general population per 1,000 live births <sup>1</sup>	Prevalence in very premature infants per 1,000 live births	Prevalence in infants born at term per 1,000 live births
Motor deficiencies	1.5 to 3	50 - 100	≤ 1.5
Visual deficiencies		10 - 40	
VA <sup>2</sup> less than 3/10	0.20 to 1.81		
Blindness	0.10 to 0.5		
Auditory deficiencies		10 - 40	
Loss fixed at 40 dB	1.1 to 1.3		
Bilateral loss greater than 70 dB	0.4 to 0.7		
Intellectual deficiencies		50 - 150	
IQ between 50 and 70	5 to 80		
IQ < 50	3.8		
Psychiatric disorders (autism and IDD)	2 to 6		

<sup>1</sup> at the age of 5 years or above; <sup>2</sup> visual acuity (in the best eye after correction)

**French data on the prevalence of severe deficiencies are similar to international data**

The information relating to infant/child handicaps in France is either taken from a permanent and continuous recording within the register framework, or *ad hoc* transversal or cohort surveys. The principal deficiencies investigated are motor, intellectual, sensorial and psychiatric.

Two registers currently exist in France - one in the Isère region and the other in the Haute-Garonne region. The Isère register (Register of infant/child handicaps and perinatal research institute, RHEOP) was created in 1991. In order to be included in this register, a child must have at least one severe, principal deficiency (defined above) and be a resident in the Isère region at 7 years of age. The main source of information is the Commission départementale de l'éducation spéciale (Special education departmental commission - SEDC). The other possible sources are the centres of early medico-social action (CAMSP) and hospital or specialist departments. The Haute-Garonne register is more recent since this was set up in 1999.

Three *ad hoc* or transversal surveys were carried out within Inserm unit 149. The first two surveys, which were conducted in 1985-86 and 1989, focused on all children presenting with severe deficiencies and born in 1972, 1976 and 1981, and whose parents were residing in one of the 14 regions covered by the survey (a total of 6,013 children between 9 and 14 years of age). The third study involved all deficient children born between 1976 and 1985 whose parents were living in three regions (Isère, Haute-Garonne and Saône-et-Loire) between 1992 and 1993 (6,174 children between 8 and 17 years of age). The SEDC proved to be the main

source of information for these surveys. A survey was also carried out involving all handicapped children born in 1984 and residing in the Lorraine region (357 children). Finally, more recently, the Handicaps, incapacities and dependence survey (HID) carried out by Insee focused on a representative sample of persons living in specialist institutions in 1998 and on a sample of persons residing in these establishments in 1999. The main aim of the survey was to describe the incapacities and their consequences. More than 1,000 children at least 15 years of age were included in each sample.

The cohort surveys include populations of children at high risk of handicap, especially the very premature infants. The first was carried out in 1985 in children born between 25 and 32 AW in the Paris region, who were monitored up until 2 years of age. The second survey was carried out on an identical population of children born in Franche-Comté in 1990-92 and followed up until 5 years of age. The third involved all of the very premature infants managed in the Fort-de-France neonatal resuscitation department that treated all children born live at less than 33 AW in Martinique between 1992 and 1995. Lastly, in 1997, all of the very premature infants born between 22 and 32 AW in 9 regions of France were included in the EPIPAGE study and followed up until 5 years of age. It was at this specific age that a full medical and psychological evaluation was organised in the centres set up for this purpose.

Currently, 9 in 1,000 infants born in the early 1990s suffer from a severe deficiency. The prevalence of motor deficiencies affects more than 3 in 1,000 children and almost 2 in 1,000 in the case of cerebral palsy. The prevalence rates for visual and auditory deficiencies range from 0.2 to 1.3 per 1,000. Finally, depending on the extend of the mental retardation, estimations for multi-handicapped children vary from 0.7 to 1.3 in every 1,000.

In the very premature infants, the first studies reported between 5 and 13% cases of cerebral motor infirmity and almost 15% of cases of moderate or severe mental retardation. More recently and focusing on a larger sample, the prevalence of cerebral palsy reached 8% at 2 years of age (EPIPAGE study).

#### **Comparison of prevalence rates of principal deficiencies in the general population and in very premature infants based on French data**

	Prevalence in the general population per 1,000 live births	Prevalence in very premature infants per 1,000 live births
Motor deficiencies	1.16 to 4.02	50 - 130
Visual deficiencies		
VA <sup>1</sup> less than 3/10	0.60	
Blindness	0.2 to 0.3	
Auditory deficiencies		
Loss fixed at 40 dB	1.1 to 1.3	
Bilateral loss greater than 70 dB	0.7 to 0.8	
Intellectual deficiencies		150
IQ between 50 and 70		
IQ < 50	1.5 to 3.5	1.5 to 3.5
Psychiatric disorders (autism and IDD)	0.5 - 1.5	

<sup>1</sup>: visual acuity (in the best eye after correction)

The Isère handicaps register shows that the number of children presenting with a severe deficiency has increased from 6.85 per 1,000 in 1980-82 to 8.85 per 1,000 in 1989-1991. This change reflects the increase in the prevalence of motor deficiencies (+26-42% for cerebral palsies) and psychiatric disorders (+90%) between 1980-1982 and 1989-1991. The marked increase in the latter could reflect changes in the way in which disorder classifications are recorded rather than a real increase in the prevalence rate. No change has been reported for intellectual, auditory and visual deficiencies.

In the very premature infants, a comparison of the results of the 1985 survey carried out in the Paris region with those of the EPIPAGE survey, which was conducted in the same region in 1997 does not reveal any change in the prevalence of motor deficiencies. None of the data allow changes in other, sensory or intellectual deficiencies to be followed up in these children.

Numerous factors are likely to influence the prevalence estimations provided by these studies. One of the crucial elements for these studies is the recording quality. As regards this particular point, the results depend on the way in which the information is collated. In the cohort studies, patients lost to follow-up were the main cause for concern. They represent between 7% and 20% of children taking part in the French studies but the published data show that they constitute a population at high risk of handicap. In the transversal surveys and the registers, the problem has been one of exhaustivity. The exhaustivity rate is 86% in the Isère register. This is difficult to evaluate in transversal surveys. These facts suggest a slight underestimation in the prevalence of deficiencies in the results produced by registered as those arising from cohort surveys.

The terms used to define a deficiency may differ depending on the studies in question. Such is the case for motor deficiencies. Cerebral motor infirmity (CMI, a French term) and cerebral palsy are two entities that match up but which cannot be superimposed on each other. However, studies recently carried out in France increasingly refer to cerebral palsy. Other phenomena may trigger differences. In particular, the degree of severity of the deficiency may vary between studies. The choice of the age at which data are registered can also affect the prevalence of handicaps. If the information is recorded at too early an age, then not all of the deficiencies, especially those of an intellectual nature, can be identified. Lastly, the quality of the data collated will play an important role. In transversal and cohort studies as well as registers, the sources of information are numerous as indeed are the number of people involved. This cannot guarantee the homogeneity of the data collection.

As regards changes over the coming years in France, two phenomena could help to reverse the trend observed to date. The first concerns the improvement in the antenatal screening of congenital deformities and a constant increase in medical involvement in pregnancies since the end of the 1980s. The second refers to the improvement in the neurological prognosis of populations at high risk of handicap such as very premature infants. This is, in fact, suggested in foreign data that confirm a stabilisation in the prevalence rate of cerebral palsy since 1990. Other phenomena could, however, counter-balance the effects of deformity screening and the management of very premature infants. In particular, if the age at maternity increases, then the number of infants born with deformities will also increase. Similarly, if prematurity continues to increase, the population of children at high risk of sequelae will also rise.

Based on the data presented above, it is interesting to estimate the annual number of children with a deficiency of perinatal origin amongst the population of children born full-term and the number of children in the premature population (also distinguishing those born very prematurely). If 1% of children between 7 and 8 years of age is considered to present with a severe deficiency, the number of these children in France can be estimated to be 7,500 (750,000 births each year) for one generation. According to the published data, the proportion of handicaps of perinatal origin varies from 15% (low estimation) to 65% (high estimation). An average estimation of 50% seems reasonable. Moreover, premature infants are considered to represent approximately 50% of children suffering from a severe deficiency.

## Estimation of the annual number of children presenting with a handicap of perinatal origin in France

	Low estimation	Average estimation	High estimation
Proportion of deficiencies of perinatal origin	15%	50%	65%
At term (50%)	560	1,875	2,435
Premature < 37 AW (50%)	560	1,875	2,435
Very premature < 32 AW (25%)	280	937	1,220
Total	1,120	3,750	4,870

Thus, in one generation, 1,120 to 4,870 children will develop a severe deficiency of perinatal origin. Half of them were born prematurely (before 37 AW) including 280 to 1,120 very premature infants (born before 32-33 AW).

### Perinatal events can be implicated in the onset of deficiencies or handicaps

The various perinatal diseases having a significant impact on the onset of handicaps or deficiencies can be classed in three major groups: prematurity and especially very early prematurity; neurological accidents in babies born full-term, mainly perinatal asphyxia; the other diseases threaten brain development.

#### Principal perinatal factors in deficiencies and handicaps

- Prematurity (sequelae associated with the gestational age, birth weight and prematurity-related complications, essentially cerebral lesions and bronchopulmonary dysplasia)
- Pre- and post-natal growth retardation
- Twin births (especially monochorial twins)
- Perinatal asphyxia
- Toxic foetopathies (alcohol and drugs, etc.)
- Perinatal infections (meningo-encephalitis)
- Various neurological accidents (neonatal stroke, trauma and haemorrhage, etc.)

Prematurity and very early prematurity in particular (birth before a gestational age of 32 weeks) has a major impact on the risk of all types of handicaps.

Evidence relating to various sequelae depends on the child's age. As a general rule, motor sequelae are diagnosed before 2 years of age and remain relatively fixed. Other disorders (learning or behavioural disorders) are detected much later at a pre-school or school age. The latter are progressive and some of them are compensated for or regress over time such as, for instance, language disorders or social behaviour disorders. However, in the case of premature infants presenting with motor sequelae, the gap in non-motor skills persists and even tends to become accentuated over time.

Different factors influence the outcome of premature infants: intrinsic factors such as gestational age, birth weight, sex, twin births or neonatal diseases (cerebral lesions and bronchopulmonary dysplasia, etc.) and environmental factors such as pre- and post-natal care or family and social environment.

In the case of very premature infants, the early use of neurological exploration techniques improves the quality of the prognosis. This is especially evident with cerebral imaging – cerebral echography, magnetic resonance imaging (MRI) – and electroencephalograms (EEGs) during the neonatal period. The clinical neuromotor examinations carried out in the first few months post-term are useful although their limitations in terms of predictive value must be remembered. These are sometimes difficult to interpret and their accessibility or quality vary from one neonatology unit to the next.

In full-term newborn infants in the event of neurological accident (perinatal anoxia, cerebrovascular accident, spontaneous intracranial or traumatic haemorrhaging and neonatal meningitis, etc.), the prognosis varies considerably depending on the nature of the accident. The best-established prognosis concerns neonatal encephalopathies, which are most generally assumed to be post-anoxic. These are manifested by a change in neurological behaviour during the first week of life. Minor encephalopathies primarily have no effect upon the child's outcome. Conversely, however, moderate and severe encephalopathies have a very important impact. Thus, in the case of moderate encephalopathy, neurological sequelae, half of which are cerebral palsies, are observed in 25% of newborn infants. The other sequelae are essentially cognitive (intellectual deficiency or language disorders). In cases of severe encephalopathies, 50% of surviving children present with neurological sequelae. Convulsions multiply the risk of motor infirmity 3-fold.

In full-term babies suffering from neonatal encephalopathy, the prognosis is based on three types of early examinations: a clinical examination during the first days of life; imaging, mainly MRI and electrophysiological examinations (EEG and/or evoked potentials). The predictive nature of these examinations, especially MRI, is described as very good in the literature (under optimal conditions). However, the experience of practitioners, who are confronted with various cases of falsely reassuring imaging, must be taken into account.

There are other perinatal risk situations, the impact of which on the onset of handicaps is more difficult to evaluate, either because they are rare or because they are combined with another risk situation (e.g. prematurity). Regardless of the prematurity and hypotrophy factors, twins, and monozygotic twins in particular, may present with clastic cerebral lesions *in utero* or during the post-natal period and deformities. The risk of lesions is all the greater when a transfuser-transfused syndrome is present.

There is a link between the presence of intra-uterine growth retardation (IUGR) and the occurrence of handicaps, regardless of the term of pregnancy. This risk is enhanced in premature cases. The prognosis appears to be chiefly conditioned by the severity of the hypotrophy. Prognostic factors in the event of IUGR are: perinatal asphyxia, gestational age, masculine sex and a small head circumference (especially the failure of the head circumference to catch up). All of these factors potentiate the risk of intellectual deficiencies in particular.

Antenatal exposure to toxic substances (alcohol) or post-natal exposure (neurotoxic and ototoxic medicinal products), post-natal malnutrition, perinatal infections, severe jaundice and severe neonatal diseases warranting a prolonged stay in an intensive care unit are also likely to promote the onset of sequelae, especially developmental problems and deafness.

All of these neonatal diseases demand prolonged follow-up, but this approach is not straightforward to arrange. A considerable difficulty for paediatric follow-up must be taken into account. Neuro-developmental sequelae are observed with higher frequency amongst families who fail to comply with follow-up schedules. At the present time in France, extended follow-up is proposed for children who are most at risk (early premature infants and neonatal encephalopathy in full-term babies). Even if it was applied, this follow-up policy would overlook more than half of the handicap-deficiencies of perinatal origin.

## Different pathological situations can be seen at the origin of perinatal events involved in the onset of deficiencies or handicaps

Different pathological situations can affect events that impact upon the onset of a deficiency or handicap such as prematurity, extensive prematurity, encephalopathy in full-term infants and other risk events.

Approximately 1/3 of prematurity is induced by medically consented and 2/3 are spontaneous. Within the scope of spontaneous prematurity, a distinction is made between premature labour on intact membranes (1/3 of premature labours) and premature labour on ruptured membranes (1/3). Variations however exist according to gestational age since the work on intact membranes affects 25% of premature labours between 27 and 32 AW, 44% between 24 and 26 AW and 64% before 24 AW.

The risk factors exposing children to moderate prematurity and extreme prematurity are the same. The link with prematurity is, nevertheless, greater for certain risk factors including the socio-economical level, living conditions (single mother, etc.), the age of the mother, the body mass index, the obstetric medical history and smoking. Intra-uterine growth retardation is also a risk factor in prematurity.

Premature labour is often associated with another complication of pregnancy: arterial hypertension (2%), *placenta praevia* (4%), retroplacental haematoma (7%) and haemorrhagic disease (16%). Placenta anomalies (*placenta praevia* and retroplacental haematoma) are frequently associated with extensive prematurity (50%) as well as infections (38%) and immunological factors (30%). Cervical incompetence (16%), maternal diseases (10%), trauma and major surgery (8%) and foetal anomalies (6%) are also related factors. These causes are interlinked in 58% of cases and isolated in 38% of cases. No factor has been identified in conjunction with prematurity in 4% of cases.

Prenatal infection is also a significant cause of preterm delivery with intact membranes. Asymptomatic, intra-amniotic infection is observed in 13% of cases involving premature investigation of intact membranes. Chorio-amnionitis will develop secondarily in 37.5% of cases. These infections are also directly involved in the onset of cerebral lesions observed in premature infants.

Twins present with an extremely high risk of prematurity and low birth weight. The risk of prematurity is increased 10-fold and the risk of weight below 2,500 grams by 11. The excess risk is observed for both moderate and extreme prematurity. Thus 10% of twins are born before 32 weeks compared with 1% of single infants. Similarly, the proportion of infants weighing below 1,000 g is 4% in twins and 0.4% in single infants. The condition of infants at birth is characterised by a higher number of stillborn infants and babies having an Apgar score of less than 8 at one minute or five minutes in the case of twins compared with single infants.

The threat of premature labour (TPL) is the primary cause of hospital admissions during pregnancy in France (38% of the 20% of pregnant women admitted to hospital).

## Various prematurity risk factors

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### Obstetric factors

- Multiple pregnancy
- Placenta praevia
- Intra-uterine growth retardation
- Congenital uterine malformation
- Cervix incompetence

### Maternal factors

- Genital, urinary and cervico-vaginal infections
- Diabetes
- Pre-eclampsia (pregnancy-related arterial hypertension)
- Rhesus allo-immunisation

### Socio-economic factors

- Primiparity; multiparity
- Unfavourable transport and working conditions
- Mother aged under 18 or over 35 years of age

### Foetal causes

- Chromosomal aberrations
- Other congenital anomalies

### Iatrogenic causes

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Neonatal encephalopathy is another risk factor affecting the onset of deficiencies and handicaps at birth. Neonatal asphyxia is, in approximately 50% of cases, a consequence of labour, otherwise it can be prevented or screened. In fact, certain cases of per-partum asphyxia may be a consequence of antepartum pathological conditions. The other diseases screened by neonatal asphyxia indicators could be exclusively pre-natal pathologies.

The entire problem stems from the major difficulty involved in identifying per-partum asphyxia in situations where the indicators are hardly specific and common to the entire perinatal period.

It is difficult to specify the relative role of various per-partum anoxia risk factors since this varies depending on the populations studied. In favoured countries, the risk situations are placental insertion anomalies, primiparity, alcohol abuse, pre-eclampsia, the masculine sex of the fetus and intra-uterine growth retardation.

## Diagnostic criteria of per-partum asphyxia

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### Essential criteria for the diagnosis of per-partum asphyxia

- Metabolic acidosis: pH < 7.00 and base deficiency  $\geq 12$  mmol/l
- Early encephalopathy in an infant born  $\geq 34$  AW
- Quadriplegic cerebral palsy or dystonia

### Criteria that overall suggest a per-partum origin

- "Sentinel" hypoxia event: uterine rupture, pro-incidence, etc.
- Sudden and prolonged deterioration in foetal heart rate, just after the event
- An Apgar score of between 0 and 6 more than 5 minutes post-partum
- Multivisceral involvement
- Modified early cerebral imaging

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As regards the impact on the approach adopted during labour, study results tend to diverge. Generally speaking there does not appear to be a routine Caesarean policy that would reduce the number of children suffering from cerebral palsy.

The risk situations for intra-uterine growth retardation are as follows: embryopathies of chromosomal, infectious, genetic and toxic nature including the “foetal alcoholisation” syndrome and foetal malnutrition. Vasculo-renal syndromes remain the main cause of hypotrophy at birth (approximately 50% of cases). Risks are also heightened by multiple pregnancies, smoking, chronic vascular diseases and certain maternal diseases. Echography plays a crucial role in the screening and diagnosis of intra-uterine growth retardation. Its sensitivity varies depending on the term. It appears to be optimal at 34 AW with sensitivity of the order of 60% for a specificity of 85%. Some authors recommend combining clinical and echographical criteria in order to improve screening and evaluation of the severity of intra-uterine growth retardation, and propose predictive scores.

## **Early screening of cerebral lesions is essential but not life-threatening**

Early screening of cerebral lesions or identification of the risks of impaired cerebral development are essential in a child. In fact, early intervention now holds an important place amongst the possibilities currently available for action.

Cerebral lesions acquired at the end of pregnancy or during the perinatal period represent the majority of lesions responsible for paediatric neurodevelopmental handicaps. Early screening can be carried out:

- before birth based on current echographist performances that reveal porencephalopathies, ischaemo-haemorrhagic strokes, microcephalies, calcifications of the cortical grey matter or basal ganglia, cytomegalovirus-induced foetal diseases in particular and ventricular dilatations;
- at birth in the presence of revealing clinical symptoms in a full-term newborn infant: early neonatal encephalopathy or isolated convulsions or hypotension;
- at birth by systematic cerebral imaging monitoring (transfontanellar ultrasonographic study or nuclear magnetic resonance imaging) of certain populations at risk: extremely early prematurity, twin pregnancies, even intra-uterine growth retardation or infants with an excessive birth weight;
- during the months following birth in the presence of early neuromotor abnormalities sometimes combined with growth retardation of the cranial perimeter during monitoring procedures in certain populations at risk during mandatory examinations performed at 9 months and 2 years in the general population.

The circumstances under which cerebral lesions are discovered vary, depending on the extent of the lesions and their topography. Their clinical expression during the immediate neonatal period is often poor when accidents have occurred at the end of pregnancy or in a premature, newborn infant. Routine cerebral imaging amongst populations at risk (twins, infants presenting with, intra-uterine growth retardation and premature babies) will reveal cerebral lesion of ischaemic and/or inflammatory origin. Sometimes, the clinical picture will be more extensive, especially in full-term infants subjected to diffuse anoxic accidents affecting not only the basal ganglia but also the grey matter. Their clinical detection will often be delayed in relation to birth because of secondary neuromotor abnormalities.

Lesion topography is, in itself, closely related to the stage of cerebral development. Thus, given the vulnerability of its white matter, very premature infants often present with a disease affecting the white matter accompanied in particular by periventricular leucomalacias or intraventricular/intra-parenchymatous haemorrhages. In full-term infants, given the functional maturity of the neurones, grey matter and grey nuclei, early neonatal

encephalopathy is evident from early neonatal encephalopathy by diffuse or localised neuronal necrosis.

The prognosis of various cerebral lesions and the importance of sequelae are directly related to the aetiology of the lesions but also to their extent and the stage of cerebral development at the time of the accident. Severe, secondary, intellectual deficiencies will rather be specific to the full-term new-born infant subjected to diffuse anoxia whereas more moderate deficiencies with praxic, memory and educational learning disorders will be observed in more premature infants. Determination of a specific prognosis is still difficult, even impossible, for several reasons. The concept of prognosis remains highly subjective. Perinatal lesions occur in a fully developing brain. Abnormal images do not inform us about subsequent cerebral functions and the brain's capacity for reorganisation. In most cases, only a risk situation, the relevance of which will vary from one individual to the next, will be apparent.

However, electroencephalograms conducted during the first week and nuclear magnetic resonance are still currently the best tools in trained hands. Anomalies in the basic tracing in full-term newborn infants and premature infants alike help with determining the prognosis. Similarly, the presence of central grey nuclei involvement on MRI will be linked with a greater risk of motor sequelae in the event of cerebro-vascular accident or diffuse anoxic attacks. The use of these tools nevertheless presents acute problems: the reduced number of EEG experts as in neonatal imaging, discordance between the abnormalities observed with transfontanel echography and MRI and a lack of MRI accessibility in certain regions.

### **Most handicaps or deficiencies of perinatal origin can be screened in the first three years**

Three types of handicap can be identified in the first three years: motor handicap, neuro-sensorial deficiencies and disorders affecting development (autism), intellectual deficiencies being identified at a later stage. Age correction in the event of prematurity must be carried out up until 2 years of age. There is frequently an additional delay with language in premature infants.

In the first months (before 6 months), severe motor handicaps of a tetraparesis nature, frequently associated with sensorial and intellectual deficiencies are highlighted by neuromotor examination, which confirms the lack of head posture, global hypotension, impaired continued vision, eating difficulties and no spontaneous motility. These children must be assessed swiftly in specialised centres in order to complete the motor, visual and auditory evaluation.

During the second six-months of life, slow progress and delayed acquisition of head and seated posture draw attention. This is precisely the context in which diagnosis is often delayed and, in the case of non-specialist doctors, warrants referral for a specialist opinion. Development consultations in the neonatology or neuropaediatric departments or even in early medico-social action centres (EMSAC) should be used as referral options in such situations. The points noted in the medical records with the examinations performed at the 4<sup>th</sup> and 9<sup>th</sup> month allow diagnosis to be approached in a precise manner.

At the end of the first year, serious and severe developmental abnormalities must be highlighted. These are mainly numerous handicaps, tetrapareses, severe diplegias and haemiplegias. If a family is given the diagnosis, the physician in question assumes that the child will be seen again on several occasions because it is essential to assess the slow advance

in order to be certain of motor involvement. In the case of intellectual deficiencies, diagnosis is very rarely made before 3 years of age.

As soon as abnormalities are observed, the children have to be referred to development specialists who will conduct a more precise semiological analysis and investigate concomitant, particularly visual disorders for motor cerebral infirmities. Other abnormalities may develop such as flexion spasms, respiratory disorders secondary to swallowing disorders and orthopaedic abnormalities.

During the second year of life, a management approach must be structured for severe motor handicaps. Other deficiencies, especially orofacial involvement with its effect on verbal language must be evaluated. Lastly, for some children, moderate motor disorders will disappear virtually completely, allowing the diagnosis of IMC to be diagnosed *a minima* or even transient neuromotor disorders.

Visual handicaps mainly affect the very premature infants who presented with retinopathy during the first weeks of life and for whom screening and follow-up must be rigorously organised as soon as they are admitted to hospital, especially if a therapeutic approach is adopted in hospital. In the advanced stages of retinopathy, 50% of infants are at risk of blindness. For others, an ophthalmological examination is urgent in the event of strabismus (squint) or nystagmus.

Auditory deficiency must be highlighted as soon as possible. The recommended age for identifying a major deficiency is 3 months in Anglo-Saxon countries. This seems early since, in some children, early auditory abnormalities can revert to normal in the second trimester of life. Babbling during the first few months does not mean that there is no deafness. In France, the diagnosis is usually made at the age of around 9 months.

Invasive developmental disorders (autism) are investigated during several consultations. Warning signs must lead to a specialist referral.

#### **Total warning signs of invasive developmental disorders**

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No babbling at 12 months

No gestures (pointing, waving "goodbye", etc.) at 12 months

No words at 16 months

No combinations of two spontaneous words (not only echoalic) at 24 months

Any loss of competence (language or social) at any age

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In the general population, medical records are inspected and if examined thoroughly should allow most deficiencies or handicaps to be highlighted. Certain items can be added: assessment of cranial sutures, opening of thumb, test for parachute reactions, analysis of the failure to adopt a seated position.

In the populations at risk, a full diagnosis is required. This involves a motility study focusing on passive, comparative, dorsal and ventral tone, active tone, anti-gravity suspension reactions, an assessment of joint amplitudes especially in the feet, anterior and lateral drawn-seated manoeuvres as described by the French School of developmental neurology. These specific neurological examinations must be carried out by doctors trained in the neuromotor examination of young children in hospital consultations specialising in high-risk children or in EMSAC possessing a screening activity. They can also be conducted by a functional rehabilitation doctor.

A simple grid together with a few diagrams should enable paediatricians to highlight more development-related disorders. The generalisation of acoustic, provoked, oto-emissions (APOE) (plus the use of ringing toys) as well as an ophthalmological examination with "baby vision" should give a more precise interpretation of sensorial deficiency.

Chronologically, at the end of the first six months of corrected age, severe and moderate deficiencies can be highlighted but still without a specific approach. The second six-month period should allow the disorders to be identified more precisely. Severe or moderate disorders will persist during the second year and may trigger the severity of certain handicaps and test for concomitant handicaps: orthopaedic, convulsions, involuntary, parasitic movements, sensory disorders and the absence of language. Follow-up for the risk population of extremely premature infants must be extended beyond 3 years as specific, cognitive difficulties are apparent only in children around 4 years of age. This follow-up procedure calls for highly specific screening techniques that are practised by neuropsychologists.

## **Early intervention is based on the concept of cerebral plasticity**

Early interventions can be defined as all of the intended prevention and treatment strategies in the first, even the second, year of life in order to promote cerebral development and optimal structuring. They are based primarily on two major axes: conventional, educational intervention strategies (neuromotor education, educational actions involving parents, etc.) and various types of neuroprotection strategies.

The early educational intervention strategies are based on the cerebral plasticity concept. These various plasticity processes are mobilised by re-educational approaches in a fully developing brain. Recent understanding of a few of the basic mechanisms of development, especially the importance of the phenomena of cell death and synaptic stabilisation and post-natal, neuronal circuits make up the theoretical and clinical basics in favour of early intervention. The capacities for motor compensation of the immature central nervous system secondary to lesions in the pre-central area are greater than those of the adult central nervous system in monkeys (Kennard principle). In children, destruction of the nervous tissue occurs in an environment that is still only slightly or not functionally specialised, unlike in the case of adults. Furthermore, the molecular and cellular response to the lesions varies with the extent of cell maturation. Under certain conditions when feasible, neural plasticity may be beneficial even if the ensuing development trajectory differs from normal. However, it can also be unsuitable if the structural connections do not permit an appropriate environmental response.

The concept of “neuroprotection”, initially reserved for molecules protecting or preventing cell death phenomena, can now be extended to all actions promoting harmonious cerebral development and preventing the onset of specific disorders affecting the latter. A distinction is made between organisational, therapeutic and environmental strategies.

On an organisational level, *in-utero* transport to allow children at risk to be born in specialist, level III establishments, has shown its impact on the reduction in early cerebral lesions. On the other hand, the effect of labour methods (pelvic or Caesarean route, before or after work) and the beneficial role of the Caesarean section have not been fully consolidated.

From a therapeutic perspective, recent animal modelling tools for paediatric cerebral lesions have led to advances in the protection of the developing brain and even in the curative treatment of acquired cerebral lesions. Numerous molecules with protective effects have thus been used against the biological phenomena involved in cell death: pro-inflammatory cytokine antagonists, membrane stabilisers, inhibitors of free radical formation, inhibitors of glutamate release, calcium channel antagonists and the NMDA glutamate receptor as well as anti-apoptotic molecules, etc. However, the difficulty in envisaging neuroprotective treatments preventing or reducing lesions in premature or full-term infants is attributed to:

the vast array of risk factors and mechanisms responsible for cerebral lesions; with no knowledge of the time (most usually antenatal) at which the deleterious cascade resulting in the cerebral lesion begins; the poor specificity of foetal stress markers (meconial fluid, anomalies in foetal heart rate) at the start of the deleterious cascade and abnormalities in adapting to extra-uterine life (Apgar score, metabolic acidosis); and finally, the difficulty in finding therapeutic agents that can be tested in clinical trials because they do not have any harmful side effects. Transfer to the clinical trial stage in human pathology is, therefore, problematic. Only some molecules or therapeutic means have been studied to date and found to offer a certain advantage: hypothermia in anoxia in full-term new-born infants, magnesium sulphate and fluoride steroids administered at the ante-natal stage and even nitric oxide at the post-natal stage in premature infants. Extensive multicentre trials using “cocktails” having several points of impact on the harmful cascade are both necessary and urgent.

Finally, the awareness of paediatricians of the role of the environment in a child’s mental and cognitive development (acquired) has allowed preventive measures to be introduced in several areas: preservation of the mother-child relationship and the processes of attachment (bonding), stress-reduction programmes in a hospital environment, “care geared towards the individual development of every child” in the neonatal intensive care unit, consideration of pain and costly education programmes for small children and taught at home in the United States.

All of these aspects confirm the belief that early intervention just like neuroprotection strategies are an essential element and must be evaluated more effectively. Furthermore, early involvement is totally justified from an ethical standpoint.

### **The highlighting of a deficiency or handicap must be accompanied by the implementation of a suitable therapeutic project**

Motor cerebral infirmity (MCI) or cerebral palsy is caused by definitive cerebral lesions that develop very early in life and are responsible for problems in the neuromotor field and language and praxis domains with deviations in neuropsychological and mental-emotional development that can also alter both actual and potential capacities in children. According to a French study, the distribution of different clinical profiles in cerebral palsy is as follows: 40% in quadriplegia, 17% in diplegia and 21% in haemiplegia.

It is important to consider these disorders in an early, continuous and multidisciplinary context. Consistent with the care and neuro-orthopaedic prevention carried out in neonatology, children must benefit from motor rehabilitation provided by a physiotherapist or psychomotor expert trained in techniques specific to young children. Depending on their wishes and potential for involvement, the parents receive practical recommendations to give their infant a proper introduction into daily life.

Consequently, child management involves a list of his/her capacities and focuses in particular on the motor, sensorial and relational potential, which will be checked and highlighted throughout growth. The therapeutic project is based on a global vision of childhood development with reference to the growth schedule.

Non-pharmacological treatment involves physiotherapy (including electrostimulation), ergotherapy and orthophony, orthetic equipment, plastered postures, technical aids and a combination of these different techniques. The scientific proof of the interest shown in physiotherapy, ergotherapy and orthophony is missing despite the fact that these are always indicated and represent an essential complement to surgery.

Conventional physiotherapy and ergotherapy are sometimes supplemented by other techniques, which are recognised to varying degree as “*patterning*”, conductive therapy, equitherapy, balneotherapy, the Votja technique, the Polish programme and segmental exclusion by induced constraint. This last technique is undergoing evaluation but initial results have already proved encouraging. Methods such as hyperbaric oxygenotherapy are still at the highly experimental stage.

For twenty years or so, the treatment of spasticity has benefited from numerous methods that are difficult to apply to children: administration of medicinal products, muscular denervation agents (botulin A toxin), intrathecal injections (baclofen) or surgery (neurectomy, rhizotomy).

The child’s progress and even difficulties are highlighted by regular clinical evaluation with reference to neuro-sensorial and psycho-behavioural development scales. The results of these evaluations are periodically presented to parents in a verbal and/or written format in an easily understandable language so as to provide the answers to the numerous questions that are asked, particularly with regard to the future.

The project, which involves medical follow-up, rehabilitation care, often devices, psychological support and rehabilitation sessions is co-ordinated by a referred physician, who possesses the necessary paediatric and/or neurology and/or rehabilitation skills. A strong family involvement is desirable, especially for the project to be set up in the first place, its practical execution and regular evaluation. Information concerning environmental adaptation and preparations for the future are provided over time with anticipation: games, home, district, school, work, leisure activities and transport. In the longer term, this support can continue with the educational project, either ordinary, managed or even specialised. Autonomy is developed for as long as possible using local educational or social equipment.

An early visual function deficiency may interfere with a child’s development and impact upon all of his/her skills, whether of a motor, cognitive or emotional nature, and thus have repercussions on school performances and both social and professional insertion. The existence of a critical visual development period occurring in the first few months of life, points in favour of early intervention in visual deficiencies in young children. In most cases, these are situations involving a risk of amblyopia, which is treatable. Uncorrected ametropies are the primary cause of strabismus and functional amblyopia. They must be controlled as quickly as possible, preferably by intervening during the sensitive period, i.e. before 1 year of age. Any anisometropia  $> 1$  dioptre, any astigmatism  $> 1.5$  dioptre, any hyperopia  $\geq 3.5$  dioptres and any myopia  $\geq 3.5$  dioptres<sup>1</sup> must be managed by optical correction involving the constant wearing of glasses and, in some cases (myopias, marked anisometropias and unilateral aphakia), contact lenses. This corrective measure must be accompanied, where necessary, with functional amblyopia treatment.

Similarly, the early intervention in strabismus and other oculomotor disorders as well as potentially related functional amblyopia, conditions the quality of the result obtained. Oculomotor disorders must be treated before a child turns 3 years of age, studies having confirmed that the earlier the alignment of both eyes is obtained (around 2 years of age), the more satisfactory the long-term binocular co-operation.

The treatment of congenital glaucoma and congenital cataract is surgical and represents a therapeutic emergency in order to limit the risks of blindness and to restore the transparency of the media to allow normal vision to develop. At the same time, the retinoblastoma must be medically controlled without delay if the child’s life is to be saved.

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<sup>1</sup> Beyond this value, myopias pose a high amblyogenic risk; however, the existence of a weaker form of myopia does not rule out the possibility of an optical correction.

The rehabilitation of low vision in paediatric ophthalmology must allow the good social integration of the child. This rehabilitation involves the participation of professionals who have benefited from specific training (specialist rehabilitation, locomotion instructors, specialists in providing assistance with living from day to day, etc.). As for the management approach adopted, it must be organised as closely as possible to suit the child's location as many towns do not have the adequate structures. Low vision rehabilitation must be structured in the form of management networks or co-ordinated care networks under medical supervision.

The early support given to a very young child (under 3 years of age) presenting with severe, auditory deficiency is based on three complementary and intrinsic principles: assistance with auditory perception (early prosthetic insertion, cochlear implants, etc.); learning to "hear" using auditory aids (early auditory rehabilitation) and assistance with implementing communication allowing the child to grasp a better understanding of the world around him/her and the demands made by others (parents, siblings, family, professionals at the establishments frequented by the child), but also to express himself/herself; the involvement of parents and other family members to help them permanently adapt to the child by introducing a suitable relational balance and rediscovering spontaneous communication. This involvement will help to prepare very young children to adapt to nursery school by proposing extended personal and individual assistance.

As regards children presenting with cognitive function deficiency (all of the mental processes involved in processing information), the aetiologies are diverse albeit unknown in a large number of cases. Former terms such as "backward" or "frail" are now firmly in the past, primarily because they posed insurmountable obstacles for any form of education or rehabilitation. However, numerous terms are currently used and refer to different, theoretical frameworks, which can have important practical consequences in terms of institution and treatment. These include: intellectual deficiency, predominantly intellectual deficiency associated with neuropsychic disorders, mental retardation (slight, moderate, severe), mental handicap, progressive disharmony and cognitive functional deficiency, etc. In France, autism still poses classification problems due to the frameworks used (concept of psychosis in the French classification system or invasive developmental disorders in the international classification system).

Raising and educating children suffering from invasive developmental disorders (including autism) requires special know-how, which is not part of the normal list of parental skills and which even goes beyond the techniques regularly taught in traditional professional training courses. The efficacy of the interventions used will depend on the availability and adequate training of competent professionals with a sound knowledge of the services that can be accessed.

Treatments for autism are largely based on the behavioural principal of operating conditioning (Lovaas method) as well as on psycho-educational and behavioural approaches centred on the acquisition of cognitive and developmental skills (TEACCH method). Both approaches are used within the perspective of educational or social normalisation. Studies applying both these approaches highlight substantial gains in the cognitive development (IQ) and speech of children suffering from autism or other invasive developmental disorders (IDD). The institution of treatment at an early age seems to be an essential condition in order to guarantee successful intervention. Regardless of the location where these programmes are carried out (either at home or in specialist centres), close co-operation between parents and professionals over a long period of time is fundamental for success. The gains obtained are usually maintained once treatment has ended. The children who make the most progress are those who have the better cognitive skills to start with.

Motor, neuro-sensorial, intellectual and psychiatric deficiencies call for an early management approach that is suitable for each situation and which is based on validated methods where possible. The various techniques have still only been assessed to a minor extent in France.

### **Early stimulation programmes have demonstrated a certain efficacy for children who are at risk of developing a deficiency**

Knowledge of the various mechanisms involved in cerebral development confirms the crucial importance of the first years of life for acquiring cognitive skills. These concepts have, however, been modulated by highlighting the process of delayed plasticity and the cognitive compensation of early deficiencies by a highly stimulating life style.

For several years now, early stimulation programmes have been devised for children who are at risk of developing a deficiency: vulnerable treatment due to the fact that they belong to an unfavourable and hardly stimulating socio-economic category; premature infants or infants with a low birth weight.

Based on studies showing that the socio-economic category was a crucial factor in child development for the first five years, programmes (*Head start programs*) were developed in the United States during the 1960s for children belonging to an unfavourable and hardly stimulating socio-economic category. Positive effects were recorded in school failure rates and, at a social level, by a reduction in the number of crimes committed. Since 1990, support has been available to parents but the additional positive effects are still unknown.

Programmes have also been launched in many countries for premature infants and infants with a low birth weight, who represent a population exposed to a high risk of developing these deficiencies. These programmes are targeted either to the child himself/herself or even to the family or combined to provide assistance for both the child and his/her family. They are initiated on leaving hospital in the transition period between being discharged and arriving home or else affect post-hospital follow-up.

The first programmes targeted towards the period of neonatal hospitalisation were based on the idea that premature infants lack stimulation and babies benefit from various forms of stimulation (multi-sensorial, tactile and physiotherapeutic) provided by nurses or ergotherapists. The results evaluated within the scope of these controlled studies appear to be positive. Other programmes aimed, on the contrary, to reducing stress and boosting children's capacities for autoregulation. These were carried out by nursing staff and gradually by parents. The results were also positive with regard to the short-term development scores. The programmes given the best evaluation are those involving both parents and children and are known as NIDCAP programmes (Newborn individualised developmental care and assessment programmes). These are individualised programmes intended for babies with a very low birth weight (less than 1,500 g) in neonatal intensive care units. The short-term results are positive (weight gain, duration of hospital stay) but are more difficult to evaluate in terms of long-term development.

Other programmes carried out during the hospital-home transition period are aimed at training parents to respond appropriately to the signs given by their child. Training is given over 7 days before discharge and then for 3 months after discharge. The beneficial effects are evident at 3 years of age and continue for up to 9 years (one single study). As for post-hospitalisation programmes, they confirm the positive effects on cognitive development and on parent-child interactions in the first two years of life, especially in cumulative risk situations (prematurity and poverty).

In the early 1990s, a longitudinal, multi-site programme on early stimulation over three years, known as the Infant health and development programme (IHDP) was launched in the United States for infants with a gestational age of less than or equal to 37 weeks and a birth weight of less than or equal to 2,500 g. The results confirm positive cognitive and behavioural effects at 24 and 36 months, which are more marked for children weighing between 2,000 and 2,500 g and for slightly more educated mothers.

Generally speaking, it is a well-known fact that combined parent-child programmes are the most useful. The effects on parent-child relationships and on the child's cognitive development are positive if stimulation is maintained. Much better results are seen at a cognitive as opposed to a motor level. These programmes are especially effective when mothers have a low level of education. They therefore compensate in part for what should have come from the family. The results show that the children benefiting from these programmes experience display less of a cognitive decline than children in control groups. This constitutes a genuine preventive action.

It remains to ascertain how risk groups should be selected considering studies where programme efficacy has been proven. A combination of prematurity and/or a low birth weight with an unfavourable socio-economic background appears to correspond to a combination of risks that clearly respond to the programmes proposed.

### **An economical screening and management approach is still barely used in decision-making**

Examination of the economic literature relating to the screening and the management of handicaps of perinatal origin is highly deceiving in more than one respect.

The published studies date back some years and essentially concern the evaluation of prematurity costs. Such paucity in the recourse to economic evaluation tools and, in particular, the total lack of any recent French study can be explained by the fact that economic analytical methodologies, if they existed, are sometimes difficult to instigate, especially due to the rate of pitfalls inherent in the cost inventory.

Cost studies provide information on the nature of financial flows used when carrying out a prevention or patient management programme. This expression of cost is generally advanced to plead in favour of the allocation of financial resources to a category of actions or health system agents. The numbering of cost can also be of a macro-economic nature and aimed at answering the following question, for example: "what is the cost to the society for prematurity and related handicaps?" A recent study carried out on this topic in California is based on a cohort of children with a low birth weight admitted to the neonatal resuscitation centre. The results clearly show the interest in preventing early premature, not only to prevent the onset of handicaps but also to avoid the very high expenditure concerning both living and deceased children.

**Cost of management in neonatal intensive care per gestational age (surviving children) (according to Gilbert et al., 2003)**

Gestational age	Cost per child (\$)	Average age of stay (days)
25	202,700	92.0
26	146,600	75.9
27	119,600	66.8
28	86,200	52.3
29	62,600	39.5
30	46,400	30.4
31	29,800	21.5
32	18,900	14.8
33	11,000	9.0
34	7,200	5.9
35	4,200	3.9
36	2,600	2.8
37	1,700	2.2
38	1,100	1.8

The cost-efficacy analyses are aimed at comparing the “profitability” of various preventive or sanitary actions from the point of view of a collective interest. They allow various strategies to be classified per order of the decreasing cost-efficacy ratio provided that the real value of the consumed resources is measured. In the cost-efficacy analyses, consequences are determined by a “physical efficacy indicator”, which can be, for instance, a biological indicator or autonomy, the number of abnormalities screened, lives saved or deaths avoided. One of the difficulties in analysing cost-efficacy is the choice of a relevant efficacy indicator to measure the performance of evaluated actions. Medical innovations increasingly interfere not only with the “quantity of life” that they can obtain but also the quality of life for the subject who will benefit from this. Certain actions can extend the life-span but at the cost of a certain morbidity or deterioration in the quality of life of these subjects during the health procedure.

The economic evaluation can thus take the form of a cost-utility approach developed essentially in Anglo-Saxon contexts. This approach weighs the physical efficacy indicator by assessing the quality of life (QALY: Quality adjusted life year). This assessment is made on a subjective basis by interviewing medical experts or target individuals of the action or general population. This is a compared evaluation of two types of treatment of retinopathy in the premature infant (cryotherapy and laser), which links to each treatment the number of years of life in good health that are saved.

**Cost per QALY of the retinopathy screening programme applicable to the premature infant (according to Javitt et al., 1993)**

Screening strategy	Cost of programme	QALY saved	Cost per QALY (\$)
Weekly	28.1	4,648	6,045
Twice-monthly	15.7	4,334	3,623
Monthly	9.7	3,899	2,488

The cost-benefit analysis is the fullest form of the economic evaluation but also the most delicate because it attempts to link an economic or monetary value to the sanitary benefits of health care actions. This approach poses methodological problems since it is necessary to

monetise the consequences of changes in health conditions such as production losses. A British study relates the additional expenditure of screening and retinopathy treatment of the premature retinopathy to costs saved by reducing the number of visual handicapped subjects whose condition must be controlled.

Nevertheless, extreme caution must be exercised when interpreting the results of all the economical studies. A comparison of the results obtained in different countries is always risky due to the differences in social protection and sickness insurance systems.

### **Certain countries have more experience than others in organising management approaches**

In Europe there is considerable heterogeneity concerning the organisation of neonatal care for extremely premature infants. In the Netherlands, Portugal and the Czech Republic, newborn infants of less than 32 weeks are managed in level III units. On the other hand, in Denmark, Northern Ireland, Germany, the United Kingdom and France, these children are also admitted to hospital in level II units. Despite a consensus of opinion regarding the need to transfer pregnant women at risk of premature labour to maternity wings linked to resuscitation and neonatal care services, almost none of the very premature French infants were *outborn* in 1996. There is still no comparative evaluation of these different organisational decisions.

NIDCAP programmes were initially developed in the United States and then in Sweden to respond to the needs of children born very prematurely in terms of stimulation and care during a critical period of development of their central nervous system. The pain and discomfort caused by care practices during hospital admission were taken into account. These programmes require changes in the organisation of care in neonatology departments and considerable rehabilitation efforts. Rehabilitation specialists going on to become development specialists are portrayed as parents' allies, bringing them confidence and skills to take care of their child. These specialists are starting to take part in research into neonatology care as members of multidisciplinary teams.

Thirty years or so ago, the United States introduced a policy to assist children with physical, mental or behavioural handicaps. Since 1990, the programme launched by the Individuals with disabilities education act (IDEA) developed a decentralised, family-orientated system of multidisciplinary care. At the heart of the system, the "*medical homes*" represent the ideal approach to regionalised management and partly correspond to the vocation of early French medico-social action centres (EMSAC). They ensure continuity of neonatal care to adolescence, identification of requirements in medical, specialised consultations and their co-ordination, interaction with school, social life and the centralisation of information on medical care and the hospital admittance of each child. The type and extent of management approaches are based on the Individualised family service plan, an individualised, family-orientated programme, which defines the most suitable form of education for each child in line with his/her development. The paediatrician at the medical device centre discusses the child's progress, treatment and clinical course with the family and helps to locate additional services for secondary prevention purposes.

A recent study that evaluated access to the early management programme (IDEA) at national level indicates that the average age at which parents report an abnormality is 7.4 months. The diagnosis is carried out on average 1.4 months later and admission to the programme occurs on average 5.2 months after diagnosis. The individualised management plan is initiated 1.7 months later at an average age of 15.7 months. Most families are satisfied with

their involvement in an early intervention programme. They report having had an opportunity to discuss matters with a health care professional and have benefited from straightforward access to services corresponding to their requirements. They positively assess the work conducted by the professionals and felt that they played a role in the decisions taken regarding their child.

In Europe, the disappearance of specialist institutions in various countries (Sweden and Italy, etc.) led to accentuating the decentralisation of skills in terms of handicap in order to promote a local management approach. In Sweden, committees have adaptation centres for handicapped children combining screening and handicap treatment resources (physiotherapy, ergotherapy and orthophony, etc.). These centres are also responsible for the pre-school insertion of handicapped children. Where possible, handicapped children with normal intellectual capacities go to a mainstream school and are assisted, if required, by personal assistance and suitable equipment. Children suffering from an intellectual handicap and possibly other functional limitations go to a "separate" school. In Italy, the law of 1977 abolished the differentiated (streamed) classes and special classes by defining flexible forms of integration in favour of teaching handicapped children at mainstream school with specialist teaching support. The right to integration is recognised throughout the mandatory school period but also in crèches and secondary schools.

The United Kingdom comes mid-way between countries that have opted to close specialist establishments and France or Belgium where there is still an important specialist education sector. The policy is directed at having a maximum number of children in ordinary establishments with the assistance, where feasible, of external specialists but this does not imply the closure of specialist establishments, which are viewed as a complement and not as an alternative to "*mainstreaming*". In the United Kingdom, a large number of autistic children are taught in an ordinary environment with specialist support.

Comparison of the different ways of organising management approaches for handicapped children carried out by the European Agency for developing the education of persons with particular requirements in various EC states is interesting from several angles. It shows that some countries have more experience than others in organising management approaches. Common points in terms of actions and programmes nevertheless warrant emphasis. Generally speaking, early intervention is recognised as essential in the same way as a complete evaluation of the child's potential is needed in order to determine the assistance required. The teams cover four key areas of early intervention: medical, psychological, educational and social. These teams consist of additional professionals, depending on requirements.

For several years now, we have witnessed a tendency to promote "developmental care" in deficient or handicapped children in an attempt to integrate them in an ordinary environment. The services in charge of "developmental care" are most often under the responsibility of the public health or social affairs "authorities" but educational skills (schooling) are more actively needed in the programmes than in the past. Amongst the points emphasised as necessitating improvement, there are practical difficulties in organising the co-ordination of services and professionals given the lack of organisational measures and strategies.

## **The involvement of families in organising developmental care is essential**

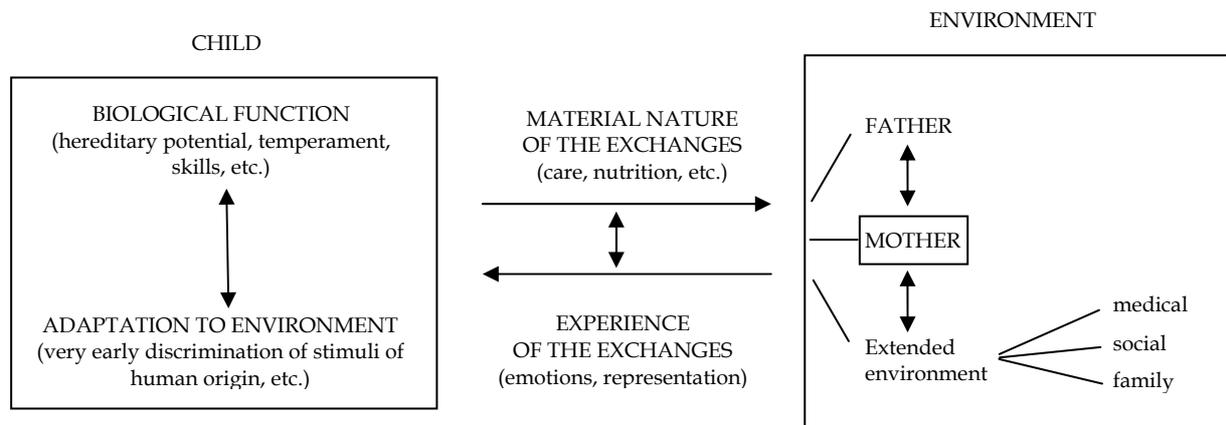
The role played by parents in the care system introduced for a very young child can be analysed at three levels:

- their involvement in the management of a child suffering from a proven or potential handicap;
- the type of support experienced by the parents, from pregnancy and subsequently, when a risk of disease is highlighted;
- the highlighting and management of their vulnerability factors in preventing certain obstetric-paediatric complications (e.g. prematurity or growth retardation) as well as the effects of perinatal measurement approaches on the forging of parent-child links.

We can rely on a simple diagram combining all of the interactive systems surrounding the newborn or unborn infant in order to integrate these three levels of reflection. These systems take into account the biological, emotional and inter-relational elements that interfere in a child's somato-psyche development.

This diagram enables a link to be made between pregnancy, the post-partum period, early infancy and also between the medical, social and psychiatric fields. The follow-up given to an extremely premature child will depend on obstetrico-paediatric co-ordination, which will have consolidated or broached parents' confidence with regard to the care system.

The interest in involving parents in the event of a handicap is aimed primarily at preventing an "additional handicap", which would come from a distorted link and poor resolution of the trauma resulting from announcing the handicap. Prospective clinical trials describing the bonding process between a mother and her premature child have shown that, in the case of a similar disease in neonatal resuscitation, the neuropsychic fate of children was strongly correlated with the quality of the emotional environment.



### Interactive systems in a child's somato-psyche structure

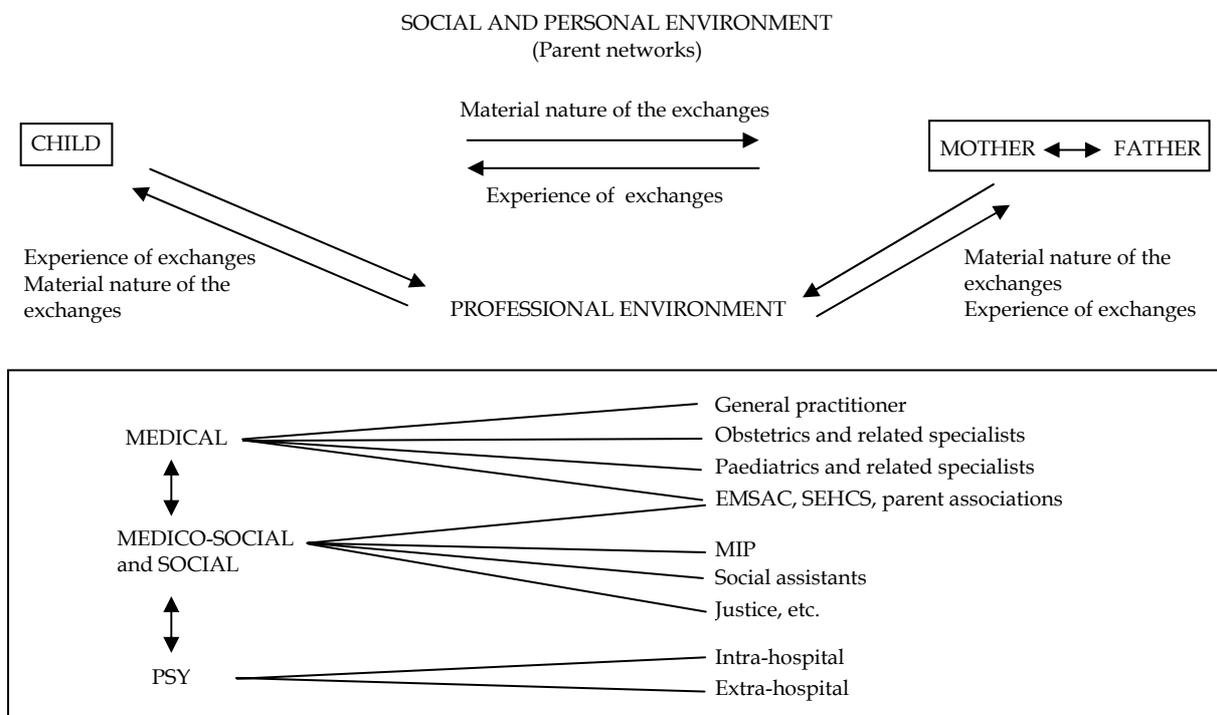
At the same time, copious amounts of published data attempt to describe the expectations of families when faced with the handicap of their child. The conditions and content of parent information become a research topic. Parents want recognition for their role and skills whilst indicating the type of support required (*counselling*): coming out of isolation, being specifically informed of the handicap and its consequences, being helped by talking to people in similar situations, being guided in finding out and accessing the various services available in administrative burdens.

The effects of parent integration in management programmes have been assessed by reducing stress, better adjustment to their child's requirements and conjugal balance. The effect of involving the father in subsequent cognitive development has been observed. These results seem to be evidence-based but this evidence was denied during the technological and institutional development period to the extent that a new type of professional has emerged:

the “insertion technician” became the parents’ mouthpiece and support when confronted with a series of services difficult to identify and rejoin.

Parental involvement is very important in certain programmes concerning autism, a condition that, in the past, was overlooked and which generated a feeling of guilt against which families strongly reacted, demanding information and respect for their role. Forms of intensive intervention integrated them as “co-educators” with positive results. Identical effects can therefore be seen between direct stimulation programmes with the child and programmes to support parent-child interaction.

Action programmes (such as Head start) launched in the United States in an attempt to reduce the dependency with regard to social assistance for families existing in the precarious situation chiefly present in intensive home visits. Their efficacy is improved when the visits begin before the birth and continue for a sufficiently long period. Antenatal visits have resulted in a reduced incidence of prematurity, growth retardation and hypertension with variants depending on the population categories, particularly ethnic origin. Better integration of the mothers has been achieved. The criteria for success in children concern the prevention of cognitive deficiencies and behavioural disorders with limited efficacy.



### Different environmental registers

EMSAC: early medico-social action centres; SEHCS: special education and home care services; MIP: maternal and infantile protection

A sociological analysis has attempted to exempt the efficacy criteria from programmes that have “worked”. These are, in fact, conditions governing participants, which appear as the most relevant indicators: autonomy, flexibility, proximity to families, a holistic approach, a feeling of competence – all of those factors offering a type of model (restoration of confidence in oneself and in others) for mothers experiencing social and emotional insecurity. The authors emphasise the interest of minor teams exempt from heavy public or private administrative burdens that divide up the action deviating from the real expectations of families, highlighting a lack of accessibility and delayed or confused responses.

Parental associations play an important role as intermediaries capable of helping to highlight the numerous specialist services, to improve the adjustment of responses and, at the same time, sharing the emotion of trauma inherent in the presence of a handicap.

In this context, clinical trials are beginning accurately to describe the interactions between professionals and parents but also within participation networks – mutual respect and complementarity seem to be a decisive factor. The notion of “alliance” between families and professionals is added to the provision of routine approaches. Thus the interesting experiences of treating and managing “in networks” drug-addicted pregnant women are manifested by a significant reduction in the incidence of prematurity, parental involvement in weaning a baby, the disappearance of authoritarian emergency placements and a decrease in frequent or repeated pregnancies. These experiences define a new clinic of “networking”, which has been made possible by the gradual decompartmentalisation of public and private, obstetric and paediatric, medical, social and psychiatric services in France in particular.

The harmonisation of interventions in the fields concerned and the early consideration of emotional vulnerability factors are important. Reflection must focus on the initial training of carers working closest to families (obstetricians, general practitioners and paediatricians) and on teaching methods that would facilitate a better mutual understanding in order to offer children and families at risk a flexible, consistent “human cover” that would continue for as long as required, regardless of the origin of the established or suspected deficiency.

## **Network organisation is essential for the success of a perinatal health policy in France**

The organisation of network care is currently presented as a model because this type of organisation is conducive to co-ordinating an action in which the potential of all those involved would be utilised: users, health care professionals, institutions and people in charge of decision-making. Various types of network already exist, some of which are formalised whilst others are not, in the field of perinatology. These networks are based on subjects, which vary in terms of specificity, interlinking various key players in the perinatology field.

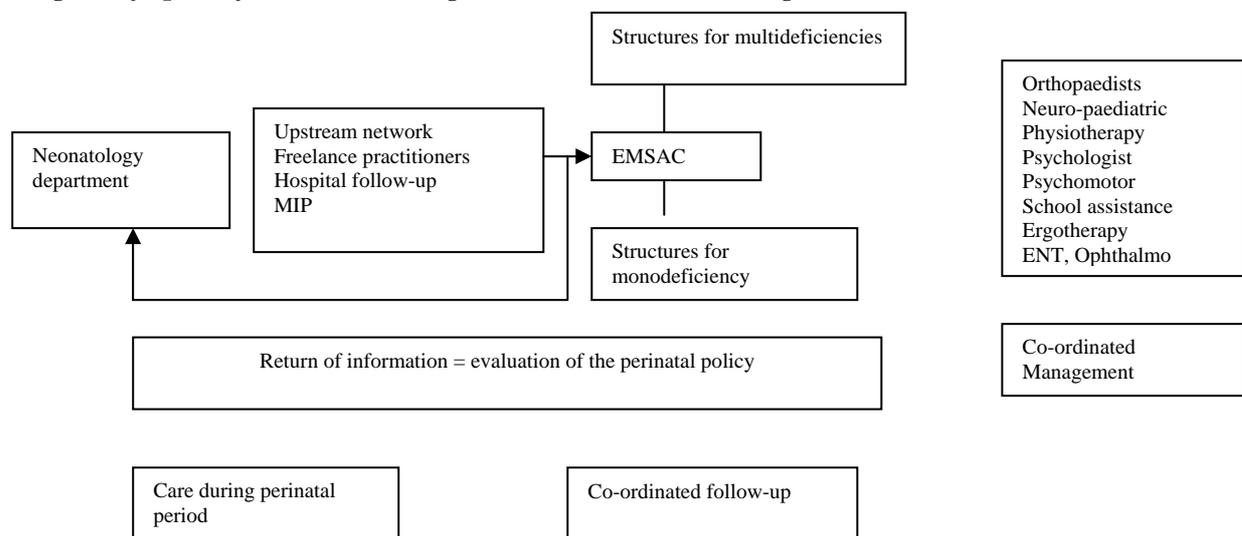
The town-hospital network for following up pregnancies in the general population links general practitioners interested in pregnancy follow-up with freelance midwives, MIP and both public and private hospital structures. These networks are directed towards more specific populations such as those that find themselves in a precarious situation. Combined town-hospital networks and inter-hospital co-ordination arrange antenatal diagnosis at regional level. The inter-hospital co-operation networks allow women to be managed in an establishment that is suitable for their medical situation. Town-hospital networks allow a return to home visits after childbirth to be organised.

Co-ordination within a “perinatal health network” of all these networks is desirable in order to maintain consistency and to allow actions of promoting the awareness of and providing information for the population to be incorporated. The aim of this type of network is to create a common culture amongst all of the participants, to make common tools available to them and to motivate them to meet the mutual shared objectives in order to improve the status of perinatal health.

Better care co-ordination will improve the health of the population. This fact has been studied chiefly in perinatology from the angle of care regionalisation, which is one of the main objectives of inter-hospital co-operation networks. Regionalisation improves perinatal health indicators in terms of perinatal mortality and neonatal morbidity. A fall in the perinatal mortality rate is especially evident from the foetal aspect. This gain is not achieved

at the cost of a deterioration in neonatal morbidity rates but, on the contrary, is accompanied by a reduction in cerebral lesions or an improvement in the development score. It is, therefore, logical to reach favourable progression for health indicators with better co-ordinated perinatal care. Care organisation upstream from birth and surrounding birth thus helps to reduce perinatal mortality. The effect of improved co-ordination of perinatal care on changes in the total number of surviving children at risk of developing a handicap is more difficult to determine. Although improved care helps to reduce the number of children at risk of developing a handicap amongst previously surviving children, it mathematically increases the total number of children at risk due to the increase in the overall number of surviving newborn infants belonging to risk populations.

A perinatal care network must involve the organisation of follow-up appointments until children at risk of developing a handicap reach school age. The parents of extremely premature infants complain of being abandoned following their discharge from the neonatology department. They are confronted by a series of professionals, who are informed to varying degree and who are competent in following up their children. The absence of a consensus opinion and references contributes to this fact. Similarly, the lack of structural co-ordination relating to specialist care in the event of single and multi deficiencies adds to misinterpretation of the health policy in the field of handicapped infancy in the opinion of the population and health care professionals. Conversely, the success of introducing perinatal care in certain regions has promoted the launch of follow-up networks known as downstream networks of perinatal care networks. The aim of such downstream networks is two-fold. The principal objective is to organise post-natal follow-up in a consistent matter in order to boost access to early management of motor, sensorial or developmental incapacities or limitations. This follow-up must allow the consequences of deficiencies to be reduced, secondary handicaps to be limited and medical error often accompanied by a feeling of parental abandonment to be avoided, whilst promoting involvement and effective, relevant assistance. The secondary aim is to carry out a long-term evaluation of the regional perinatal policy and thus of the perinatal care network. This evaluation is essential. Care practices change very quickly without having been assessed in the long term.



### Implementation of downstream network guiding high-risk patients towards suitable structures for early management

The organisation of a downstream network is based essentially on the principle of improved co-ordination of professionals and care structures. The setting up of a co-ordination cell, the creation of a common follow-up tool in the form of a common dossier, paper and

computerised on a regional server, the organisation of training to use this tool and recourse to specialist consultations are the principles means required to create such a network. The evaluation must be two-fold: quantitative and qualitative. The quantitative evaluation is relatively easy: percentage of children included and followed up until 5 years of age. The qualitative evaluation is trickier to carry out: parent satisfaction, number of children highlighted by the follow-up network and improvement of the quality of life for children receiving early management. The evaluation of networks implemented should lead to the conclusion as to whether there is a need to generalise the experiments underway.

## **Clarification and the co-ordination of institutional responsibilities that would facilitate improved management**

Although the decentralisation laws have allocated in their principles clear-cut fields of competence for each major hospital/institution, the State and general councils retain decisive skills when it comes to managing and developing the procedure for the management of handicapped people. Although this overlapping of skills is well known for the management of handicapped adults, it is less so for children under 6 years of age.

Early infancy is the period during which handicaps of perinatal origin are going to be recognised. Furthermore, some of these will not be detected before 6 years of age. Every infant is reliant on the people around him/her and it is only over time that a gap in their learning ability in relation to that of the majority of children of the same age will appear. This delay in the time taken for clinical signs to become apparent will lead to the diagnosis and justify the 20 mandatory examinations carried out in children within the maternal and infantile protection protocol (MIP).

During this period, the parents of children in whom a deficiency is expected have a choice in the approach and practitioners that they are going to use and consult, as with any other disease. EMSACs, which have a screening and family guidance mission, have entrusted the re-habilitation and care section covered by sickness insurance to general councils such as the MIP.

The State, which guarantees national solidarity, has retained the responsibility for recognising the handicap and allocating financial aid. It provides the supervision for all medico-social establishments and guides children through the various structures and establishments, apart from the EMSACs.

As regards the local management of the dossiers of handicapped children under 20 years of age, the State has entrusted the exercising of its skills to departmental commissions, the special education departmental commissions (SEDCs). These commissions recognise the handicap and assess the level of incapacity entitling sufferers to the ASE (allocation of special education) and its complements, allocate invalidity cards, direct children over 3 years of age to nurseries or medico-social structures and authorise, from birth if necessary, the intervention of care at home (special education services and care at home, SESCH). Whereas MIP regulations and actions are directed towards all children, without exception, the SEDCs must be contacted in order to be able to intervene, and they are generally approved by the parents.

All the methods for welcoming young infants, such as crèches, day nurseries or nursery assistants are placed under the responsibility of general councils, even when places specifically reserved for handicapped children actually exist. Admissions are carried out according to the methods specific to each administrative body without any need for approval by the SEDCs. The decree dated 1<sup>st</sup> August 2000, which gives parents the option to keep a

child in a crèche up until 6 years of age, and the FAF circular (Family Allocations Fund), which excludes contracts for investing in establishments that would not be able to receive handicapped children, must facilitate the admission of the latter and keep them in infant care establishments until they begin mainstream school or enter a socio-educational establishment coming under State jurisdiction.

Even if these transfers of competences follow a relatively clear logic, perception of the boundary marking out the respective fields of the two institutions often proves to be unclear to the extent that Social Security (sickness insurance and family allocations) is one of the main financiers regardless of the institution concerned. Numerous reports, including that of the Accounts Court, have stressed the confusion that arises in the role of institutional participants involved in handicap policies since the decentralisation laws came into force. Some authors conclude that even if these fragmented responsibilities, which generate joint financing, can lead certain players to deny their specific skills or not to recognise those of another.

It is a well-recognised saying that, “any handicap before 20 years of age is up to the State”, forgetting that the prevention, screening of handicaps before 6 years of age, their subsequent management by EMSACs and the methods of hosting young infants are departmental skills. Given this fact, all the readily accessible statistical data concerning the management of childhood handicaps are limited to SEDCs and to SE enquiries (specialist education), which ignore the EMSACs located outside the State’s area of competence. To use these statistical data for children of pre-school age (under 6 years of age) comes back to ignoring those for whom the handicap has not been the subject of administrative recognition.

The lack of statistical French data concerning handicaps in very young infants has led the legislator to introduce in 1973, during the first perinatal plan, infant/child health certificates to organise the collection and processing of data and to provide finance.

These certificates must be completed by the doctor who examined the child during the first 8 days of life and at both 9 months and 24 months. The same doctor is legally obliged to send this certificate within a period of 8 days and in a confidential envelope to the doctor in charge of the MIP service in the child’s home department. These mandatory investigations are carried out in full and, in order to examine the child and complete and send the certificate, general practitioners, like paediatricians, are given a paediatric fixed price, which amounts to 5 € per assessment. This fixed fee is also integrally reimbursed by sickness insurance.

These certificates are processed in each department and the database thus composed is placed under the responsibility of the MIP doctor. The processing of health certificates, data analysis and the annual transmission of results to the Ministry for Health fall within the realm of the general councils. The regrouping of data transmitted by the departments to the Ministry for Health must result in the creation of a national database.

More than 30 years after the introduction of health certificates, although the procedures are ironed out and the finances exist, the CNIL is never opposed to the computer processing of data for statistical purposes, the weaknesses of the system cannot be overlooked. There are still no reliable data regarding the perinatal period and infant/child handicaps. The quality procedure for completing these certificates is sub-standard and the quantity received by doctors responsible for MIP services is low (approximately 70% for those conducted at 9 months and 24 months), which does not mean that these assessments have not been carried out or indeed that they have been carried out incorrectly. One of the causes of this situation is that certain doctors refuse to give basic medical data to medico-social services that use such information for the purposes of individual prevention by selecting so-called “at risk” children based on medical or social criteria that do not always match.

Furthermore, there is no listing such as a SE enquiry that would identify the number of handicapped children admitted to establishments that fall under the jurisdiction of the general councils (EMSAC methods of receiving children).

The decentralisation laws have blurred the vision regarding regulations underlying the management policy for taking very young, handicapped children into care. This sharing of skills has led to the increase in a standard phenomenon in France, which, regardless of the underlying disease, involves emphasising the “care” section to the detriment of prevention and screening. This vision of things is hampered by the fact that the only national statistics that can be readily accessed concern establishments and services that fall under the jurisdiction of the State. Furthermore, the lack of reliable epidemiological data both at a national and a departmental level, makes it difficult to estimate requirements. The same applies when it comes to assessing the offer available for the management of handicaps of perinatal origin in children of pre-school age.

## Recommendations

The prevalence of severe deficiencies or handicaps is estimated at 1%, based on all of the studies. This leads to the assumption that, every year in France, 7,500 new cases of severe deficiencies appear, 50% of which are of perinatal origin. Approximately, it can be said that half of the cases related to children born prematurely. Severe deficiencies or handicaps are more frequent in premature cases and in very early premature infants in particular, but it should not be forgotten that premature births represent only between 1 and 2% of all births. Nevertheless, a number of actions such as *in-utero* transfer – which is aimed at allowing births in specialist establishments – neuroprotective, therapeutic strategies or even “individualised developmental care” are guided towards preventing prematurity on the one hand and sequelae on the other hand. Carried out in neonatal intensive care units, these latter integrate stimulations through the mother-child relationship and reduce stressful situations for the premature or low birth weight infants in order to promote their development.

The organisation of screening and the management of deficiencies and handicaps during the first three years of a child’s life, which differ slightly from one country to the next, must be able to improve in France. Several points must be emphasised: rigorous screening is required for early management adapted to each child based on validated methods. Family involvement helps to improve programme efficacy. Improved co-ordination of professionals and structures will enable networks to be set up that can better respond to family requirements.

The work of an expert group leads to proposed recommendations regarding the informing and training of professionals and families for early screening and the promotion of greater co-ordination of different players in prevention and care programmes. This work has also highlighted a certain number of gaps in terms of epidemiological knowledge in France, a lack of standardisation recording the collation of data detrimental to comparisons between different countries and the need to continue multidisciplinary research into devices and practices and to evaluate programmes.

### Boosting awareness and training

#### TO INFORM PROFESSIONALS AND COUPLES ABOUT RISK SITUATIONS

The measures taken within the scope of the national perinatal programme initiated during the 1970s and the perinatal programme (1993-2000) (personnel training, prenatal and confinement monitoring, establishment networking, guidance for women whose pregnancies are at risk and the circulation of new treatments) have allowed considerable progress to be made in terms of reducing the perinatal mortality rate, which fell from 21 per 1,000 births in 1972 to 7 per 1,000 births in 1998. Despite this progress, the overall prevalence of deficiencies and handicaps of perinatal origin does not appear to have fallen either in France or Europe. The increase in prematurity and very early prematurity since the end of the 1980s (combined with the increase in the proportion of children with very low birth weights) could be one of the explanations. However, it must not be forgotten that 50% of deficiencies and handicaps

occur in full-term infants. Unfavourable socio-economic conditions, exposure to various toxic substances (alcohol, smoking and other products, etc.) during the pre-conception period and throughout pregnancy constitute recognised risk factors of prematurity and handicap.

The expert group recommends ensuring early and free access, thanks to the overall care system for populations in risk situations, often without social cover. The group advocates introducing a care protocol as soon as the pregnancy is announced. This medical follow-up of pregnancy must include a detailed interview focusing on the psycho-social aspects and screening of risk factors at the start of pregnancy. Health professionals should benefit from the support of various networks and associations working in conjunction with ill-favoured populations.

Multiple pregnancies represent a risk of prematurity and therefore of handicap. Over the last twenty years, the incidence of multiple pregnancies (twin and triplet births) has increased due to the increase in age of women on entering maternity and the development of sterility treatments. The expert group recommends informing women of child-bearing age, and couples in particular, of the risks associated with the first late pregnancies, i.e. after 35 years of age. It suggests denouncing the triviality of recourse to ovarian hyperstimulation, improving the incorporation of this practice and monitoring the application in the “good practices” as part of the medical assistance given to procreation.

#### **TRAINING HEALTH CARE PROFESSIONALS INVOLVED IN BIRTHS AND YOUNG INFANTS**

Health care professionals involved in births and who deal with following up young children have to face the difficulty of announcing a handicap to a family. They are not usually trained to deal with such situations. The new medical course involves teaching how to support families in these situations. The expert group recommends continuous, multidisciplinary training for all professionals involved in births and young children. Thus, carers who are closest to the families (general practitioners, obstetricians, paediatricians, nursery nurses and rehabilitation staff) could offer these families a consistent, human environment.

#### **DEVELOP THE TRAINING OF HEALTH CARE PROFESSIONALS FOR IMPROVED SCREENING OF NEURO-DEVELOPMENTAL DISORDERS IN YOUNG CHILDREN**

The screening of neuro-developmental disorders during the first three years of life could be improved provided that certain knowledge regarding the neuro-development of the young child was mastered. In cases of very early prematurity, the corrected age has to be taken into account in order to assess children’s development up to 2 years of age and to monitor the clinical course during all clinical and neurological examinations (tone, consciousness, motility, cranial perimeter and visual and auditory examinations) from the first year of life so as to establish the extent of the progress being made. For children over 3 years of age, professionals specialising in the care of young infants must recognise cognitive disorders as early as possible and especially at the start of nursery.

The expert group recommends training health care professionals (general practitioners, paediatricians, physiotherapists, orthophonists, psycho-motility specialists and nursery nurses, etc.) in the techniques and scheduling of early screening for neuro-development abnormalities. The creation of a common screening platform at regional level should assist this training programme. It also suggests that these professionals be made more aware of the problems in the mother-child relationship during maternity. The professionals must recommend a management approach that is suitable for parents when an abnormality is

identified. The expert group advises making neonatologists more aware of economic studies that can highlight the interest of incorporating early screening and prevention in the management strategies.

## Preventing risk situations

### TO PREVENT CEREBRAL LESIONS IN EXTREMELY PREMATURE AND FULL-TERM INFANTS

The beneficial impact of *in-utero* transfer on mortality and morbidity rates in the short term has been clearly demonstrated for children with a gestational age of less than 30 weeks. The expert group recommends promoting *in-utero* transfer to level III maternity care for children with a gestational age of less than 30 weeks. Consequently, it suggests a good distribution of level III maternity care in relation to population areas and consistent distribution of the care available by promoting the mother and child relationship. The expert group draws attention to the situation in the Paris region, which is particularly difficult in terms of managing this distribution of available care. It should be noted that, in this region, 50% of the very early premature births occur in level I or II maternity establishments.

Antenatal corticosteroid therapy has had a beneficial effect on severe forms of intraventricular (IVH) and intra-parenchymatous haemorrhages (IPH) and other frequently encountered inflammatory diseases in very premature infants. The results tend to vary with regard to the effect on periventricular leucomalacia, which confirms a different effect depending on whether dexamethasone or betamethasone has been administered. The expert group recommends prescribing an antenatal corticosteroid therapy – with betamethasone – in cases at risk of premature labour between the 24<sup>th</sup> and 34<sup>th</sup> week of gestation. It suggests detailed usage avoiding repeated courses of treatment as studies have revealed a toxic effect if more than 2 courses are administered. It is therefore important to carry out a benefit/risk analysis and to evaluate the immediate neonatal benefit.

In full-term infants after encephalopathy, moderate, early hypothermia could involve treatment preventing the onset of cerebral lesions. Positive results have been recorded in Australian studies. The expert group recommends paying attention to tests currently underway, the preliminary results of which are conclusive.

Longitudinal studies demonstrate the overlapping role of peri- and post-natal health-related incidents and the environment on the fate of infants. The expert group stresses the importance of maintaining, in a neonatal resuscitation unit, the mother-child relationship promoting the bonding process, adapting “developmental care” programmes for each child, taking into account the pain that premature infants may experience and reducing the phenomena of stress that are present to a considerable extent in a hospital setting. It recommends developing parental guidance programmes and helping parents as well as providing education programmes for affected families that can be watched at home.

### PREVENTING PREMATURITY AND VERY EARLY PREMATURITY

The prevention of very early prematurity would be a means of reducing the prevalence of deficiencies and handicaps. The limited time use (48 hours) of tocolysis in a parental format can, in cases of imminent premature labour (IPL), allow the *in-utero* transfer of the child and

introduction of corticosteroid therapy to promote lung maturation and limit the risks of cerebral lesions.

The highlighting of urinary or vaginal infections and the institution of antibiotic therapy can prevent premature labour. The expert group recommends that professionals refer to the good practices used in gynaecology and obstetrics.

## Early screening

### TO IMPROVE SCREENING DURING THE FIRST TWO YEARS IN THE GENERAL POPULATION

The highlighting of neurological development is ensured by a certain number of items in the medical records, which, once recorded, provides a framework for detecting developmental disorders. In the general population without a specific risk situation, abnormal development is highlighted by tests carried out by the midwife at birth (Apgar test, weighing and cranial perimeter) and by a full examination comprising a neurological examination (tone, behaviour, reaction to noise and continuous vision), which is performed by the paediatrician when mother and baby are discharged from the maternity ward. Routine examinations (at 4, 9 and 24 months) allow motility, prehension, language and the social interaction of young children to be assessed. Despite the individual variability in neurological development, an upper limit of acquired motor skills has been established for full-term infants: supporting the head at over 4 months, remaining seated at no later than 9 months and walking no later than 18 months. The expert group recommends developing a formal guide to infant examination to be attached to the medical records.

The expert group also suggests that every professional involved with young children should refer any child presenting with the following clinical signs to a neuropaediatric department or an early medico-social action centre (EMAS): abnormal development of the cranial perimeter, swollen head after 5 months, persistence of hypotension in the lower limbs after 5 months, the lack of voluntary prehension at 6-7 months, no seated posture at 10 months, failure to search for a hidden object at 12 months and a lack of walking after 20 months.

It also draws attention to definitive warning signs of invasive developmental disorders (including autism): no babbling at 12 months, no gestures (waving goodbye with the hand) at 12 months, no words at 16 months, no spontaneous combination of two words (not only echoes) at 24 months, regardless of any loss of skill (language or social) at any age. The expert group recommends implementing the necessary measures to reduce the delay between the first highlighted clinical signs of autism and diagnosis, and subsequently between diagnosis and management. It proposes that resource centres be developed at regional or departmental level (information, evaluation and networking of various services, etc.).

The future of a child with impaired hearing depends on the early identification of the auditory deficiency and its management since the auditory perception of language during the first six months of life conditions subsequent language development. There is a great deal of interest in screening at national level because neonatal deafness is more frequent than other abnormalities that are already subject to routine screening (congenital abnormalities such as hypothyroidism, phenylketonuria or drepanocytosis). The expert group recommends investigating the introduction of routine screening of auditory disorders as early as possible. The American Paediatric Academy suggests that this should be carried out before the age of 3 months.

An early visual function deficiency can affect the development of all of the child's motor, cognitive and emotional skills. Amblyopia is reversible with treatment but only during a period starting with the first few months of life and terminating around 5 or 7 years of age. Diagnosis and treatment must therefore be introduced at an early stage and definitely before the child reaches 3 years of age in order to obtain maximum restoration of visual function. The screening of visual abnormalities is covered within the scope of the general follow-up procedure initiated for children. The expert group recommends investigating the feasibility of a routine assessment at 9 months (orthoptic examinations, refraction measurement and testing for any organic abnormalities). Visual acuity can be evaluated from 30 months onwards by means of morphoscopic tests (image recognition) since the child is capable of responding verbally. The expert group suggests that this measure be taken during the child's first year in nursery or maternal and infantile protection centre to manage functional amblyopias that have previously gone undetected.

#### **IMPLEMENTATION OF CONDITIONS FOR THE EARLY SCREENING OF DEFICIENCIES OR HANDICAPS IN CHILD POPULATIONS AT RISK DURING THE FIRST YEARS OF LIFE**

The following child populations are considered to be at high risk of developing a deficiency or handicap of perinatal origin: very premature infants, full-term infants with neonatal encephalopathy or another perinatal neurological accident (occurring during the first week of life), children having presented with life-threatening diseases close to birth, children presenting with cerebral lesions screened during the perinatal period and those with cranio-facial deformities, severe jaundice or severe heart disease.

Repeated examinations are essential during the first two years in order to highlight a developmental disorder in high-risk children. In addition to the standard follow-up, the expert group recommends 6 to 7 consultations, 4 to 5 of which should take place during the first year. If an abnormality is detected, the number of consultations must be increased in order to propose an appropriate management programme.

Clinical examination of the child should allow motor disorders to be detected at full term (gestational age of 40 weeks), at 4 months and at 8-9 months. Cerebral motor infirmity (CMI or cerebral palsy) is diagnosed when a series of clinical signs are present: haemiplegia and swallowing disorders are generally recognised towards the sixth month by parents before the professionals. The typical clinical picture of CMI includes problems in maintaining the posture of the trunk and head whilst stiffness of the lower limbs is perhaps less typical. The clinical course during the second year will confirm the diagnosis by differentiating between spastic and dyskinetic attacks and by specifying the affected areas. Premature infants generally have a lower performance rating regarding 4 motor items (motor capacities, fine motor skills, global motility and visuo-motor integration). The expert group recommends relying on these four items to test for motor deficiencies in premature children.

As regards visual deficiencies, paediatricians and ophthalmologists must ensure regular and well coded ophthalmological follow-up for very early premature infants in view of screening for retinopathy, which poses the risk of blindness. The expert group suggests regular follow-up during the neonatal period for all premature infants weighing less than 1,500 g or born at less than 32 weeks. Following the neonatal period, the examination includes evaluation of external motility, measurement of refraction under cycloplegia, examination of the back of the eye and visual acuity tests suitable for the child's age. If the premature infant presents with retinopathy, follow-up must continue up to 8 years of age. Moreover, very early prematurity is a substantial source of visual problems (25 to 50% of very early premature infants present with visual disorders), which are further enhanced in cases of retinopathy.

With regard to the screening of auditory problems, the expert group recommends using provoked acoustic oto-emissions (PAOE) before the child is discharged from the neonatology unit. If the examination gives an abnormal reading, then the early auditory evoked potentials (EAEP) test must be carried out at the age of 3 months.

Children at high risk require close follow-up involving a multidisciplinary team trained in detecting neuro-sensorial and cognitive disorders and aware of the psychological difficulties experienced by families. The expert group recommends regular follow-up at home during the first months of life and continued follow-up thereafter through consultations carried out twice a year, even in the absence of clinical signs.

The populations of children considered to be at risk of developing a deficiency or handicap to less of a degree are children born moderately premature (33-36 weeks of gestation), children who were hypotrophic at birth, twins, children presenting with materno-foetal disease or a prenatal addiction and children presenting with an excessive birth weight. Follow-up is particularly recommended for these children in the event of a concomitant disease or if the children in question come from an unfavourable social background. In addition to standard, routine examinations, the experts recommend a specific follow-up for these children at least once a year up until 8 years of age because the cognitive or behavioural sequelae may appear at school age.

In order to reach child populations at risk of developing a deficiency or handicap, medical follow-up should be accompanied by a social network since populations that are the most exposed are often the most difficult to monitor or to maintain in care networks.

#### **TO DEVELOP AND STANDARDIZE TOOLS FOR THE SCREENING OF EARLY LESIONS**

It is important to have tools available for the screening or even the selection of children at risk to allow neurological follow-up during young infancy since lesions can be identified not only at birth but also slightly later during follow-up procedures.

In the event of neonatal, so-called anoxic encephalopathy, isolated neonatal convulsions indicative of cerebro-vascular accident or hypotonia revealing an antenatal or a periparturient accident, the expert group emphasises the interest of electroencephalograms (EEGs) to confirm the functional status of the brain allowing intellectual prognosis to be made mid-term. However, even in the case of a good prognosis, regular follow-up up to school age is recommended because learning deficiencies may not be detected by standard exploration during the neonatal period.

Overall, echography is a good tool for screening cerebral lesions in premature children but its predictive value is limited. In premature and especially very premature infants, transfontanel echography and/or nuclear magnetic resonance imaging (MRI) allows intracranial haemorrhages and periventricular leucomalacias responsible for motor infirmities to be screened.

The off-spring of twin (and multiple) pregnancies or born with intra-uterine growth retardation or an excessive weight can also benefit from screening via cerebral imaging if the slightest doubt arises. Recent studies in fact show that these children are at risk of developing cerebral motor infirmity, intellectual insufficiency or learning difficulties. The expert group recommends using all of the imaging resources for improved follow-up of children at risk. The use of telemedicine can promote the reading of examinations (MRI, echography and EEG).

## Management

### TO GENERATE “DEVELOPMENTAL CARE” PROGRAMMES

The management of affected children or children at high risk of deficiency or handicap is gradually organised around the concept of early intervention, which covers a set of measures geared towards children and their families in order to stimulate a child’s sensori-motor, emotional, social and intellectual development. “Developmental care” programmes have been launched and evaluated for different child populations.

In the neonatology department, NIDCAP (*Newborn individualised developmental care and assessment program*) programmes are applied to babies with a very low birth weight (less than 1,500 g). They propose the organisation of appropriate care, which prevents excess stress for the baby. These programmes have a short-term beneficial effect on weight gain and the duration of the hospital stay, and some studies also show the effects on motor, cognitive and behavioural development after a 3-year follow-up.

To expedite the return home, programmes are aimed at training parents at how to respond in a suitable manner to the signs given by their child. The training course is given over a period of 7 days prior to discharge and then for 3 months thereafter. The beneficial effects are evident at 3 years of age and continue up until the child reaches 9 years of age.

Early intervention or “developmental care” programmes put into practice after a hospital stay have shown the positive effects on cognitive development and parent-child interactions during the first two years, particularly in cumulative risk situations (prematurity and poverty). All of the methods used within the scope of these programmes in different countries are aimed at catching up with child development in order to promote the child’s integration in a mainstream school environment.

For autistic children, early, intensive programmes based on the behavioural principle of operating conditioning (Lovaas technique) or on psycho-educational and behavioural approaches focused on the acquisition of cognitive and developmental competence (TEACCH method) have proven their efficacy. In fact, they allow the integration of some children in a school environment with or without special assistance. Results following the evaluation of all these programmes show that parental involvement is essential for greater efficacy.

The expert group recommends that “developmental programmes” be applied early to children presenting with deficiencies or a handicap or who are at a high risk of developing these (premature infants or infants with a low birth weight). They emphasise the interest of these programmes chiefly in children born into families experiencing financial difficulties, who are in a precarious position or are isolated and who cannot ensure themselves that the child will be adequately stimulated.

### TO PROMOTE CO-ORDINATION OF THE INTERVENING SERVICES IN THE MANAGEMENT APPROACH

The early management of a child presenting with a deficiency or handicap warrants the intervention of multidisciplinary teams covering the medical, psychological, educational and social fields. Generally speaking, early interventions is recognised as essential as indeed is a precise and complete evaluation of the child’s potential before deciding what assistance is required. It focuses on informing the parents at a very early stage of the treatment and care

structures and on assistance and recourse to the special education departmental commission (SEDC) and the family allocation fund. Where possible, it is preferable for the child to remain within his/her family context. Early guidance towards a motor rehabilitation institute (day care) or to SESHC (out-patient basis) is an option that is available after notifying the SEDC. The expert group stresses the importance of developing an adequate number of early medico-social action centres (EMSAs) and of improving the way in which these centres function.

The expert group suggests early management of the handicap within the scope of an individualised management plan. The aim of this approach is to ensure that each child is given the appropriate developmental care and rehabilitation/education allowing him/her to develop to his/her full potential so that, if possible, the child in question can be integrated as soon as possible in a mainstream school environment. The expert group consequently proposes developing crèches and playgrounds as mixed reception structures accepting a fraction of different children and creating care and living locations.

A programme (entitled, for instance, "Handi-reception") designed along the lines of the Handicol' programme launched in the educational field in 1999, could be introduced. It should include all of the institutions including ordinary establishments (crèches, playgrounds and nursery schools). Its basic principle would be to emphasise regional, co-ordinated and network-structured policies based on the logic of management continuity and individualised approaches. The expert group recommends developing such programmes and promoting organisational strategies ensuring the co-ordination of services and different participants in order to prevent allocating spread-out contacts to families.

#### TO VALIDATE EXPERIENCES IN PERINATAL CARE NETWORKS

Numerous experiments of "network" care organisation are currently underway in France within the field of perinatology. These experiments take different forms: the town-hospital network, which ensures the follow-up of pregnancy in the general population or, more precisely, in populations in a precarious situation; inter-hospital co-operation networks (regionalisation of care) enabling women to be monitored and treated in an establishment best suited to their medical situation and town-hospital networks that arrange the return home following confinement. The co-ordination of all these networks within a "perinatal health network" appears desirable as this would make common tools available in an attempt to improve perinatal health. The setting up of perinatal care networks should help to create follow-up networks and to train professionals involved with young infants.

The expert group recommends evaluating the experiences acquired through networks currently underway and to promote their development in other regions to create effective screening and follow-up networking procedures. To improve management, the expert group proposes a centrifuge structure of equipment for assistance with, at regional level, the recognition of centres of expertise possessing multi-disciplinary technical platforms (for instance, within small day-care hospital units and full hospital admission, follow-up and rehabilitation care, physical medicine and paediatric rehabilitation at university medical centres) in conjunction with a network corresponding to a medico-social and educational network at local level linked with regional medical and paramedical specialists.

## Promoting research

### TO IMPROVE KNOWLEDGE OF THE PREVALENCE OF CHILD HANDICAP IN FRANCE AND ITS TRENDS COMPARED WITH OTHER EUROPEAN COUNTRIES

The two registers currently in existence (Isère and Haute-Garonne) are inadequate to investigate the prevalence of handicaps and deficiencies in France because they cover only 24,000 births (out of an annual total of 750,000 births). They do not allow studies to be carried out by sub-groups according to the level of risk: populations at low risk (full-term infants), at moderate risk (moderately premature infants born 33-36 weeks AW – amenorrhoea weeks – or born at full-term with growth retardation) and those at high risk (very premature infants born 22-32 AW). They do not, therefore, carry out case-control studies. The expert group recommends developing a sufficient number of registers (one for 75,000 births) throughout the country in order to ensure a truly representative level.

The expert group suggests defining the types of deficiencies or handicaps to be recorded according to international classifications and to homogenise this collated information at SEDC level. If the optimal age of recording for cerebral palsies is 5 years, it seems to be appropriate to continue the recording up to 7 years of age because certain deficiencies can only become apparent at a later stage.

To improve data collection, the expert group advocates using health certificates, which have been intended to be a source of information for more than 30 years, and to create a young infancy-specific database on the structures and types of management approach adopted by all hospitals/institutions and finance sources.

It seems important to investigate the prevalence of deficiencies in each of the sub-populations in association with trends in events during the perinatal period: medical interruptions of pregnancy, perinatal and neonatal mortality rates and levels of prematurity and early maturity. The expert group recommends developing, in the mid-term, a monitoring system combining obstetric and birth-related data (gestational age, birth weight, single and multiple pregnancies, etc.) with data entered in the registers.

When analysing data, it is important to take into consideration the medical practices that differ from one country to another in order to compare prevalence rates at European level. The expert group advises registering all pregnancies over 22 weeks of amenorrhoea and to introduce European registers taking changes in practices and diagnostic classifications into account.

The expert group recommends facilitating long-term follow-up of very premature infants in current cohort surveys (EPIPAGE cohort) to introduce new population surveys and to develop methodologies suitable for studying populations at intermediate risk by extending the follow-up period to beyond 5 years.

### TO DEVELOP RESEARCH INTO THE AETIOLOGY OF DEFICIENCIES OR HANDICAPS OF PERINATAL ORIGIN

Deficiencies or handicaps are considered perinatal if they occur between 22 weeks of amenorrhoea and the 8<sup>th</sup> or 28<sup>th</sup> day of life. Several different types of event may be involved during this period. The resulting deficiency may pass unnoticed for several months or even several years, hence the difficulty in establishing a causal relationship between an event and a deficiency or handicap. The expert group draws attention to the interest of a birth cohort

such as the EDEN cohort (study of pre- and post-natal factors in a child's development and health) to identify a certain number of risk factors, risk situations and predictive values of deficiencies and handicaps.

New tools are likely to screen early lesions in risk populations. They could help to boost an understanding of the physiopathological mechanisms causing developmental disorders and propose new prognostic markers. The expert group recommends assessing the interest of new tools, especially in functional cerebral imaging and electroencephalography: diffusion MRI, positron emission tomography, spectral EEG analysis and motility analysis in altimetry...

#### **TO DEVELOP STUDIES ON THE PHYSIOPATHOGENY OF CEREBRAL LESIONS AND PROTECTIVE TREATMENTS**

The physiopathogeny of cerebral lesions has still not been fully elucidated. It is now admitted that cerebral damage is the consequence of the combined action of several factors involving exogenic aggression (such as hypoxo-ischaemic aggression or inflammatory or infectious aggressions in premature infants) with abnormal endogenous mechanisms of cerebral development (deficits in terms of growth factors or the weakness of anti-oxidizing systems in premature infants, etc.).

The expert group recommends continuing research into various phases involved in the formation of cerebral lesions in an attempt to identify a therapeutic window that would allow preventive action of cellular lesions. Although numerous molecules aimed at protection have been tested in animal models to counteract the onset of cerebral lesions, clinical trials have proved difficult to implement. The expert group suggests encouraging clinical trials on the most promising and least harmful molecules (magnesium sulphate, erythropoietin, corticosteroids and anti-oxidizing agents, etc.) and other treatments (hypothermia).

#### **TO COMPARE THE VARIOUS EARLY INTERVENTION STRATEGIES IN FRANCE**

It is important to manage deficiencies and handicaps right from the outset adopting a continuous and multidisciplinary approach. The management procedures are based on a wide range of techniques, not all of which have been evaluated. The management techniques must be adapted to suit each of the deficiencies (motor, sensorial, intellectual and mental). These different approaches are generally used from the perspective of reverting school or social life to normal. Regardless of the location where the programmes are carried out (at home, school or in specialist centres), close co-operation between parents and professionals is a condition in order to ensure success.

The expert group recommends assessing and comparing various types of early intervention available in France in the motor, neuro-sensorial, cognitive and educational domains and development of quality of life scales for the child and his/her family (including siblings).

#### **TO DEVELOP MULTIDISCIPLINARY RESEARCH INTO THE DEVICES AVAILABLE AND PRACTICES USED FOR CHILDREN BETWEEN 0 AND 6 YEARS OF AGE IN A HANDICAP SITUATION**

In France, we do not have an overall view of the population of children in a handicap situation or health problem between 0 to 3 years of age and 3 to 6 years of age. It is *a fortiori*

virtually impossible to find out the distribution of children in these age brackets falling into the category of cognitive functional deficiencies. Experts are largely of the same opinion in their desire to carry out routine surveys supported by a computerised approach, allowing them to take stock of the various early management procedures used for handicapped children. The spread of structures and collection of information does not currently provide a satisfactory global analysis or the ability precisely to define the requirements with regard to structures. Such assessments should be carried out, bypassing existing institutional divisions since they also concern the sanitary as well as the medico-social and educational sectors.

The expert group recommends promoting research in order to have a vision of all the various management approaches for handicapped children between the ages of 0 to 3 years and 0 to 6 years. Such research into handicap and incapacities could be developed within the framework of a national research institute.