Shaken baby syndrome and prevention programs

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Summary
Shaken Baby Syndrome often occurs after shaking in response to crying bouts of infants; which are a source of frustration and anger for parents. It results in serious health problems in the infants such as intracranial and retinal bleeding, brain injury and even death. The syndrome can be prevented by providing training to parents, especially in the early postnatal period and increasing awareness about the dangers of shaking. This review aims to draw attention to shaken baby syndrome which is largely unrecognized in our country and to emphasize the importance of the prevention programs. (Turk Arch Ped 2012; 47: 150-156)

Key words: Prevention program, shaken baby syndrome, training program, Turkey

Introduction

The definitions of “head trauma related to abuse” or “non-accidental head injury” (NAHI) are used in addition to “battered child syndrome”, “shaken baby” or “shaken baby syndrome” (SBS) to describe physical abuse resulting in brain and skull damage in children (1).

Non-accidental head injury is most commonly observed in the first year of life and is the most common unnatural cause of death in infancy (2,3). SBS which is defined as a subgroup and the most commonly observed type of non-accidental head injury is closed head trauma which occurs as a result of recurrent acceleration and deceleration mechanism in young children. Typical clinical characteristics include diffuse brain damage, retinal bleeding and subdural bleeding (SDB) and sometimes posterior rib and metaphysial bone fractures can also be observed (1,2).

History

Child abuse was defined by Tardieu (4) (a French doctor) for the first time in modern medicine in the second half of the 19th century. Tardieu reported a blood layer was present on the surface of the brain in a child who was exposed to abuse and described all types of child abuse and reported that it could lead to physical and emotional disorders (4,5).

John Caffey, an American pediatric radiologist, reported babies with long bone fractures and subdural bleeding. Caffey did not thought that these findings could be related to abuse, but reported that they could have occured as a result of small accidents. In 1962, Kempe (6) reported his observations about “battered baby syndrome” and this publication is considered as the first scientific article about child abuse.

Ommaya (7) performed experimental studies on the mechanisms of damage by applying whip movement on Rhesus monkeys. This study was the first study showing the mechanism of shaking experimentally.

Caffey described the association of subdural bleeding, retinal bleeding and fractures in long bones which occured without any history of trauma or disease. Ludwig and Warman (8) used the term “battered baby syndrome” for the first time.

Epidemiology

Use of different definitions and lack of collection of the data in one center make reliable determination of the frequency of shaken baby syndrome difficult (1,6,9,10).
In our country, lack of consideration of shaken baby syndrome by healthcare workers and omitting autopsy because of accepting mortal cases as death related to accidents can be added to the difficulties experienced in the whole world in the diagnosis of shaken baby syndrome (11).

More than 90% of severe head traumas in infancy and 80% of pediatric deaths due to head trauma below the age of two arise from non-accidental head injuries (3,12).

In limited epidemiologic studies, the frequency of shaken baby syndrome has been reported to be 14-33.8/100 000 in children below the age of one (13,14). It has been reported that these figures are below the actual predicted frequency because of lack of diagnosis or erroneous diagnosis and are strongly related to low socioeconomic status (13).

It is reported that 1200-1500 children are exposed to shaken baby syndrome and 300 children die because of shaken baby syndrome each year in USA according to 2005 data (15).

There is only one multi-center study reporting the frequency of non-accidental head injury in children below the age of 3 in Turkey (11). Children below the age of 36 months who were admitted to the hospital because of subdural bleeding (SDB), subarachnoid bleeding (SAB), skull fracture, coma, retinal bleeding and change in consciousness were included in this study retrospectively. 48 of the cases were determined to be accidents. 7% were determined to be possible abuse and 22% were determined to be abuse. In 23% of the cases the cause could not be determined.

**Risk factors**

Child abuse is affected by multiple factors. It occurs as a result of interaction of the risks related to the baby, risks related to the caretaker and social and environmental risks.

**Risk factors related to the baby:** Victims of shaken baby syndrome are generally below the age of one. It is found most frequently in infants between the ages of 2.5 months and 4 months (8,16,17,18). Cases have been reported in infants at the age up to 6 weeks and in children older than 5 years (19).

Disabled children constitute 14% of the children who are proved to be abused. The rate of neglect has been reported to be 1.8 fold higher in disabled children compared to children without disability. The rate of physical abuse has been reported to be 1.6 fold higher and the rate of sexual abuse has been reported to be 2.2 fold higher (20,21).

Male infants, multiple pregnancy, premature babies, babies with a low birth weight and babies with weak bonding between the baby and the caretaker carry a higher risk (18,20,22,23).

It has been found that the frequency of shaken baby syndrome increases in parallel to the increase in the frequency of crying of the baby. Therefore, crying is accepted to be the triggering mechanism for SBS (19,24). Persistent crying of the baby leads to anxiety in the parents who can not understand why the baby is crying and do not know how to behave. This is the basic trigger of violence and increases anger. Increase in anger leads to loss of control. This stress can grow suddenly because of inadequate social support and shaking occurs with loss of control (25).

Anatomic and physiologic properties of babies predispose to damage by shaking. Although babies have a small body volume, head to body ratio is high. Therefore, it is difficult for the weak neck muscles to control the head during shaking and the head flies back and forth with angels reaching 240 degrees. Open sutures and a large subarachnoid space cause the head to move further inside the skull and subdural bleeding via rupture of suspensory veins below the dura. Incomplete myelinization and high content of water in the brain cause neuroaxonal damage (2,8,20,26).

**Risk factors related to the parents and caretaker:** Common properties of SBS exploiters have been determined, though not defined precisely (Table 1). The most common exploiters are men namely the biological father, the boyfriend of the mother and stepfather (16,17,20,22,27).

**Environmental and social risk factors:** Parents whose children are hospitalized have many risk factors in terms of SBS because of increased stress, anxiety, hostility, depression, loss of control and disordered system. Parents who have ill and disabled children are under emotional and financial pressure in terms of daily and social life, medical needs and financial expenses (20) (Table 1).

While some studies have reported that this syndrome is observed more frequently with low socioeconomic level, some have reported that it is observed with an equal frequency in all social categories (1,13,21,28,29).

<table>
<thead>
<tr>
<th>Table 1. Risk factors related to the exploiter and environmental risk factors in shaken baby syndrome</th>
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<tbody>
<tr>
<td>Male gender</td>
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<tr>
<td>Divorced or single parent</td>
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<tr>
<td>Low education level (&lt; 12 years)</td>
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<tr>
<td>Unemployed parents</td>
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<td>Being parent at a young age&lt;24 years</td>
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<tr>
<td>Habit of consuming drug-alcohol</td>
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<td>Personal and familial history of abuse</td>
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<td>Being a caretaker because of necessity</td>
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<tr>
<td>Unrealistic expectations of the baby</td>
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<td>Unhappy marriage</td>
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<tr>
<td>Poor family relations</td>
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<td>Social and cultural behaviour pattern</td>
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</table>
Pathogenesis

Since all samples and theories have limitations, the mechanism of injury has not been fully elucidated. Injury occurs by rapid and recurrent flexion, extension and rotation movements of the head and the neck which stays stable relative to the head (7,26,30). The exploiter usually holds the baby’s chest, arms and shoulders or rarely legs and shakes the baby strongly. The speed of shaking is generally 2-4 times per second, but can also be 5-20 times per second. After shaking the baby the exploiter may throw the baby towards a surface (20,31).

In the light of the study of Ommaya where whip movement was applied in Rhesus monkeys, Guthkelch reported that the reason for brain damage was the whip movement during shaking and bilateral SDB occurred as a result of rupture of bridge veins arising from rotational force of shaking (32).

While some studies have reported that shaking alone can lead to findings in children, others have claimed that shaking alone can not lead to SBS findings and impact head trauma is efficient in severe brain damage (26,27).

During shaking the brain moves inside the skull and dura which are relatively more stable. The time difference between the movement of the skull and the movement of its content causes direct trauma, damage and rupture of blood vessels. Damage to blood vessels leads to intracranial bleeding. Subdural bleeding is the most common intracranial finding in SBS. Typical marked blood collections of 2-15 ml occur between the subdural hemispheres (2,20,21,26). Increased intracranial pressure occurs secondary to subdural bleeding, brain edema and brain damage (4,20,26).

It has been reported that the first brain damage in a shaken baby is caused by hypoxia and this leads to brain edema and increased intracranial pressure in order (33,34). In addition, oxidative stress, inflammation and damage triggered by iron lead to injury, because antioxidant systems have not matured fully in babies (35,36).

It is possible that the force which causes brain damage leads to damage in the child’s neck which has a weaker structure (1). In severe and fatal cases, cervical spinal damage may occur without spinal fracture and this may be one of the factors in the pathogenesis (30).

The force which creates shaking causes stretching and rupture of the retinal vessels. The reason of retinal bleedings include acceleration-deceleration mechanism, brain edema, increased intracranial pressure due to compression of SDB and increased intrathoracic pressure (23,37,38).

Generally, neurologic damage and death occur because of ischemia and uncontrolled brain edema (34,39). The degree of brain damage depends on the severity of shaking, the force applied, the presence of throwing and the time to reach medical treatment (20,26).

Clinical picture

The findings of shaken baby syndrome are variable. Severe and rapidly developing findings of head trauma may occur or nonspecific mild findings may be observed (8,20,29).

When shaking is not forceful enough to lead to exitus or neurologic findings, the victim may present with feeding difficulty, vomiting, somnolence and restlessness lasting for days or weeks. These non-specific findings may be underestimated by physicians or may be defined as viral disease, colic and nutritional deficiency (40). In such mild cases, the findings improve until the actual cause is determined. In some cases, the diagnosis is made when the baby is reabused or chronic subdural bleeding findings are observed (for example: enlargement of head circumference) (5).

After unconsciousness occurs, the caretaker who has shaken the baby may put the baby on the bed with the expectation that he/she will recover after a while. This causes loss of chance for early treatment. Generally, the caretaker will not give information which will explain the situation and state that he/she has found the baby in this state (3,12,40).

Common signs include unexplained tendency to sleep and respiratory arrest (47%) (Table 2) (8,16,20).

On physical examination, frequently no external finding can be found. Signs should be investigated and recorded and visible findings should absolutely be photographed (2,9,16).

Retinal bleedings with no history of trauma indicate SBS. In 70-90% of the patients, unilateral or bilateral retinal bleedings are present (16,23,38,41,42,43). In a meta-analysis, it was found that retinal bleedings which developed related to non-accidental head injury were typically diffuse, symmetrical on both sides and localized in many layers posteriorly or adjacent to the ora serrata. Optic nevus sheath hemorrhage has a sensitivity of 72% and a specificity of 71% in non-accidental head injury (43). Studies have reported that there is a relation between the extend of the retinal bleeding and the severity of trauma and mortality (3,16,38).

Subdural hemorrhage and diffuse subarachnoid hemorrhage which develop as a result of rupture of bridge veins are frequently observed findings in babies with non-accidental head injury (8,26).

<table>
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<th>Table 2. Common findings in shaken baby syndrome</th>
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<td>Malnutrition</td>
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<td>Growth retardation</td>
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<td>Vomiting</td>
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<td>Respiratory disorders</td>
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<tr>
<td>Respiratory arrest</td>
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<tr>
<td>Weakness</td>
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<td>Decreased muscle tonus</td>
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Diagnosis

Generally, history related to abuse cannot be taken. The history told by the caretaker usually includes “shaking the baby to bring him/her to life” or falling in the playground, from the bed or from the sofa or injury by accident. This is a clue to be suspicious of abuse (1,8,26). When taking the history especially history of falling (“what was the child doing?” “How did the child fall, from where did the child fall, from what level did the child fall?”, “what did you do when the child fell?”), the caretaker, the relation between the caretaker and the child, previous traumas and the medical history of the child should be interrogated. In addition, factors which might have triggered abuse (the child’s crying behavior, presence of colic, vaccination history, toilet education) should also be interrogated (1).

Since the history of shaking can generally not be taken, a detailed physical examination and diagnostic tests are very important for the diagnosis (8,20). Multidicipliner approach is more beneficial compared to standard care in the diagnosis (1).

Since ophthalmological findings are important in the diagnosis the examination should be performed by experienced specialists (pediatric ophthalmologist, pediatric neurologist) using appropriate devices and dilating the pupils (41,42,43). It has been reported that a false negative result can be obtained with a rate of 13% in ophthalmological examination performed by other clinicians (18).

Radiologic bone screening should be performed in children younger than 24 months, if physical abuse is suspected. Findings of injury which are obscure initially, periosteal seperations and greenstick fracture may not be visible in the early period. Therefore, follow-up bone scanning may be appropriate, if there is clinical suspicion (1,20,44).

Brain imaging should be done in all infants and children in whom non-accidental head injury is suspected. Brain imaging is recommended in all infants younger than one year old who are suspected to be exposed to abuse, who have face injury, rib fracture or multiple fractures and in all infants younger than 6 months old who have any evidence of physical abuse even if there is no clinical finding (1).

Computerized tomography (CT) is rather sensitive in determining intracranial disorders and is generally the first radiologic test performed. Direct cranial x-rays are considered only when CT cannot be performed (3). Computed tomography is preferred over magnetic resonance imaging (MRI) because of non-invasiveness, low cost and higher availability. The age of the damage, fractures, severe brain edema, subdural hemorrhage, subarachnoid hemorrhage, diffuse extra-axial hemorrhages and mass effect can be observed efficiently on CT (1,44).

Magnetic resonance imaging is a diagnostic test which supplements CT. It is more sensitive in terms of showing soft tissues and it demonstrates intraparanchimal hemorrhage, early brain edema and brain damage more clearly and distinctly (44,45). However, sedation which is needed for shooting is harmful in patients who have a tendency to neurologic deterioration (20). When CT cannot explain the clinical findings fully or obscure findings are observed on CT despite neurologic findings, MRI may be needed (45).

Diffusion imaging is a new and valuable technique in the evaluation of brain damage related to abuse. It shows ischemia which is the basic mechanism of damage especially in SBS. However, CT and MRI are still preferred primarily for the present time (44,46).

Differential Diagnosis

Since the findings of SBS are not specific, many conditions should be considered in the differential diagnosis (Table 3) (3,8).

It is important to differentiate non-accidental head injury (NAHI) form accidental head injury (AHI). Infants are younger in NAHI compared to AHI and have previous medical problems (18,39,47,48). While subdural hemorrhage is found with a rate of 8% and subarachnoid hemorrhage is found with a rate of 2% in falls from a high level accidentally when the distance is below 120 cm, retinal hemorrhage has never been observed. In NAHI, subdural hemorrhage, subarachnoid hemorrhage and retinal hemorrhage were found with a significantly higher rate (39).

In a metaanalysis, it was found that respiratory arrest and retinal hemorrhage were highly determinative findings for NAHI, rib fracture, seizures, long bone fractures and skull fractures were observed with a higher rate in NAHI compared to AHI, but the difference was not statistically significant. Since the positive predictive value of respiratory arrest is very high, it is recommended to be recorded in all cases of head trauma (18).

Measurement of markers indicating brain damage in serum (neuron-specific enolase, S100B, myelin basic protein) may be beneficial in the differentiation of NAHI and AHI (49). These markers increase in a shorter time in NAHI compared to AHI and have previous medical problems (NAHI) form accidental head injury (AHI). Infants are younger in NAHI compared to AHI and have previous medical problems (18,39,47,48). While subdural hemorrhage is found with a rate of 8% and subarachnoid hemorrhage is found with a rate of 2% in falls from a high level accidentally when the distance is below 120 cm, retinal hemorrhage has never been observed. In NAHI, subdural hemorrhage, subarachnoid hemorrhage and retinal hemorrhage were found with a significantly higher rate (39).

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Since subdural hemorrhage and retinal hemorrhages can be observed in bleeding disorders, prothrombin time, partial thromboplastin time, thrombin time, complete blood count, fibrinogen and fibrin degradation products should be tested in patients in whom head trauma related to abuse is considered (1,3).

<table>
<thead>
<tr>
<th>Table 3. Diseases which should be considered in the differential diagnosis</th>
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<tr>
<td>Sepsis</td>
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<tr>
<td>Meningitis, meningocoecephalitis</td>
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<tr>
<td>Congenital heart disease</td>
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<tr>
<td>Metabolic diseases</td>
</tr>
<tr>
<td>Accidental head injury</td>
</tr>
<tr>
<td>Conditions with seizures</td>
</tr>
<tr>
<td>Viral diseases</td>
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Subdural hemorrhage and retinal hemorrhages may be observed in glutaric acidemia type 1 which is a metabolic disease. Differential diagnosis is made by other clinical findings and observation of findings in attacks in these patients (50,51). Rarely, retinal hemorrhages may be observed in patients with galactosemia and bone disorders and subdural hemorrhage may be observed in Menkes syndrome; differential diagnosis is made by other clinical findings and laboratory tests (3). Atypical fractures are observed in osteogenesis imperfecta type 1-4; the diagnosis is made by presence of familial history, blue sclera and genetic tests (3,52).

**Prognosis**

12-35% of SBS cases result in mortality; there are studies which report a mortality rate up to 35% (1,2,9,16,53).

Damage due to shaking causes mortality or persistent neurologic damage; less than 35% of the victims are found to be normal (1,16,33,48,53,54). In survivors, blindness and vision disorder develop in 30-65%, spastic paralysis or severe motor disorder develop in 30-60%, epileptic seizures develop in 30% and speech disorders develop in 64% (9,16,53). Microcephaly, stable encephalopathy, deafness, chronic subdural fluid collection, enlargement in the ventricles and brain reduction may also be observed (45,55).

In ¼ of the victims who have no finding initially, severe disorders may arise after a long period (5,8,39,48). Attention disorders, memory problems and learning difficulties may arise after the child starts to go to school. Behavioral problems are observed with a rate of 52% and usually occur in the 2nd-3rd year of life (53).

The factors which determine persistent neurologic damage have been reported to include retinal hemorrhage, intracranial lesion, enlargement of head circumference and brain reduction. In addition, persistent neurologic damage was found to be related to younger age, prolongation of consciousness state, child trauma score and Glaskow coma score, subdural hemorrhage, decreased pressure in the brain/decreased intravascular pressure and presence of brain edema (16,53,56).

**Prevention**

Sequelae which occur after the children are shaken are frequently irreversible and even may result in death. Therefore, application of prevention programs and preventing abuse before it occurs are accepted as the primary approach.

It has been reported that 18 000-70 000 dollars are spent in the first admission of children who are exposed to non-accidental head injuries, 300 000 dollars are spent for annual medical expenses and 1 million dollars are spent for life-long care. With an efficient protection program the lives of many children would be saved and disabilities would be prevented. In addition, economic benefit will be provided for the community (10,57).

Caffey emphasized the importance of protection in 1972 for the first time. He reported that the brain would be damaged if the parents shook their babies, they should never shake their babies and in this way they could protect the organs and brain and even the lives of their young children.

Many parents do not have information about the sensitivity of the brain of an infant and the harms of shaking. In the study performed by Showers et al., it was reported that 25-50% of high-school students and the parent who had children or who would have children in the near future did not know shaking caused brain damage or mortality in infants (10). Protection can be provided with giving information and support to the parents in this aspect each time medical care is given.

In addition, education of healthcare workers in terms of SBS mechanism, risk factors, sequelae and protection methods is important in terms of diagnosis and providing notice (58).

**Protection Programs:** Raising awareness in the whole community should be the primary target. There are classically three steps in protection of abuse:

1. **Tertiary protection** methods include efforts to diminish the damage, prevent recurrence, treat and cure the child when abuse has occurred. It may be that 46% of the children with a diagnosis of shaken baby syndrome were exposed to abuse before the diagnosis (58,59). Definition of the first abuse and treatment and rehabilitation of the exploiters in addition to the victims are mandatory to prevent abuse in the future (1).

2. In **secondary protection** methods, families carrying high risk in terms of abuse should be defined. In this way, the requirements of the family can be satisfied or the children can be placed in a care center. When these families are known by healthcare workers, they can be directed to institutions which can give support and efforts can be made to decrease the risks. Efforts can also be made to increase the social support of these families. A strong social support system is efficient to decrease violence even if risk factors are present in the family (21).

The actual prevention method is **primary protection.** With this method abuse is prevented before it occurs and the parents are supported throughout the childhood. The recommended methods include prenatal and postnatal supportive programs, education related to child development, home visits, education for coping with stress and education of ways for coping with anger and fatigue which trigger shaking the baby (2). In this section, prevention programs used in the world will be discussed in detail and their significance in primary prevention will be emphasized.

A few prevention programs are applied by giving postnatal primary prevention education to parents in the whole country (10,57,60,61,62).

It is appropriate that the caretaker is actively involved in SBS prevention program. It is important to tell the parents that their feeling of inadequacy and fatigue is normal and it is important to encourage them. The caretaker should be given advises about himself/herself and he/she should be recommended to pay attention to his/her rest and sleep, plan...
daily works, exercise and spare time for themselves separate from the baby. In addition, written information is recommended for other people in the house (20).

Currently, healthcare workers give information to parents about SBS at prenatal visits and postnatally before discharge in developed countries (58). Another method is to give education to individuals who get in contact with babies in infant care centers, school courses and social workshops (20,58). House visits is the other method which is used in SBS prevention programs for families with low income level who have risk factors (55).

Inclusion of SBS education into the education program given while babies are being discharged is recommended, since it is the only opportunity to give education to the father, the mother’s boyfriend and other men living in the house who are the most common exploiters related to this subject (20,22,27). It is important to explain that crying is a normal part of development in infants to all people who take care of babies, to teach the methods to cope with crying babies and to soothe them and give information about the harms of shaking (10,20,57,60).

SBS prevention programs are being implemented in developed countries. Organization of these programs started in 1974 in USA and “Shaken Baby Syndrome National Center” was instituted in 1990. In this program, all patients who had newborn babies were given information about SBS by nurses immediately after birth in the hospital (15). As a result of this education programs head traumas were reported to be decreased by 47% in New York in a period of three years. In this study, it was reported that 10 dollars and approximately 15 minutes were spent for each baby and it was cost-effective (57).

Showers et al.(10) gave a card including information about babies’ crying, soothing methods and the harms of shaking together with the birth report to mothers who gave birth in Ohio during discharge. More than 75% of the mothers found this information beneficial, 57% reported that they obtained more information about the harms of shaking. After this study handbills including soothing methods and methods to cope with anger were arranged in different languages and in a comprehensible way and were started to be used in 50 states in USA and programs giving information about SBS were arranged for radio and TV (58).

‘The Period of PURPLE Crying’ SBS protection program is being implemented in 49 states in USA and in 8 states in Canada and ‘Love Me…Never Shake Me’ education program is being implemented in Ohio (15,60,62). In these programs, education films, handbills and remindful tools giving information about crying periods of babies, the fact that babies can cry without any reason, methods to soothe babies and anger control for parents are given.

With a protection program which started to be implemented in 2004 in Australia early family education is given with the help of tools including animation films, posters and leaflets and supportive programs especially for new parents are pursued (63). The aim of this education is to develop the methods of to cope with stress and to educate parents about the harms of shaking using a friendly language. Here, the target population includes the community, new parents, future parents, childminders, school children and other family members. “Shaking your baby is just not the deal” is used as the main motto.

In Turkey, child abuse has been brought to agenda especially in the last 10 years and an awareness about the possibility that this problem has reached serious dimensions in our country has started to be established. The sensitivity of the media about this subject has increased, some non-governmental organizations have started training activities and the government has also started activities. However, SBS as a subtype of abuse is known with a very low rate and only limited number of studies have been conducted. In the literature, only a few case reports are found related to our country. An unpublished study in which non-accidental head injuries presented to different hospitals were compiled was presented in an international congress (11). An organized prevention program is not available as far as we know. It seems to be necessary to give trainings to healthcare workers and collect data to recognize overlooked cases in our country and to initiate preventive activities with widespread education programs for the community considering this important issue of pediatric health and development. Establishing a new program appropriate for our country taking example by the programs which have been shown to be efficient abroad will provide prevention of disability and mortality for many infants and contribute to their health.

References


